POST GRADUATE LECTURES
ON
ORTHOPEDIC DIAGNOSIS
AND
INDICATIONS

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AND
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By

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VOLUME IV

SECTION A ARTHRITIS AND DISEASES OF MUSCLES, BURSAE TENDONS AND FASCIAE

SECTION B DEFICIENCY AND DEGENERATIVE DISEASES OF THE LOCOMOTOR SYSTEM



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Section A

ARTHRITIS AND DISEASES OF MUSCLES, BURSAE, TENDONS AND FASCIAE

Lecture I

ON CHRONIC NONSPECIFIC ARTHRITIS

I ORIENTATION

THE TERM "nonspecific" merely indicates that as yet no specific agency has been established as the cause of this type of arthritis There are a number of factors capable of producing identical pathological changes, but there is no single one for which the pathological picture is pathognomonic This is in contrast to the so-called specific arthritis, such as the tuberculous, gonorrheal, or pyogenic, where the immediate cause of the disease is unmistakably identified with the responsible organism

Since we know that an identical morphological picture of nonspecific arthritis can be produced by a variety of agencies, we must relinquish the idea of an etiological classification and be content with grouping cases on purely patho logical or morphological grounds From the morphological point of view, a primary division can be made into two great types of nonspecific arthritis The so called *atrophic* and *hypertrophic* types

The atrophic group is also referred to as Type I, and the hypertrophic, as Type II This division is justified by essential pathological differences which exist between the two groups In the atrophic type of arthritis, the primary changes are synovial, in the hypertrophic type, they are osteocartilaginous

Any further subdivisions can be made on clinical grounds only For instance, in the primary synovial or atrophic arthritis of Type I one may distinguish an infectious, a metabolic, and an endocrine group as clinical entities A similar clinical subdivision can be made for the second group of nonspecific arthritis or the hypertrophic arthritis of Type II Here one can distinguish between traumatic, degenerative, senescent, and endocrine types So we see that in the present light of our knowledge only the primary subdivision into the two great groups can be made on morphological grounds, while all further subdivisions are based merely on clinical manifestations

II THE PATHOGENESIS OF THE ATROPHIC TYPE OF NONSPECIFIC ARTHRITIS

It would seem contradictory to speak of the pathogenesis of nonspecific arthritis when the immediate causes of the disease have not been identified As long as one considers the bacteriological or toxic invaders as the sole cause of the disease, the objection would seem valid On the other hand it cannot be emphasized too strongly that these extraneous agents are not unconditionally pathogenic Their disease producing effect is possible only if the organism is especially susceptible to it

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One can never arrive at a proper concept of nonspecific arthritis if one looks at it merely in the light of the invader. It is at least of equal importance to investigate the reasons why a particular individual is or may become susceptible to the disease. Quite obviously there must exist a general immunity against arthritis, whatever its cause may be, for the simple reason that the causes are ubiquitous, and the disease is selective. We may say the same of tuberculosis, where the chances for infection are universal and the infection itself is the exception rather than the rule. It seems logical, therefore, to realize first of all that the individual propensity or allergy is the most important single factor in the production of the nonspecific arthritis.

Let us take, for instance, the case of arthritis caused by infection of the upper air passages such as the tonsils or sinuses. The incidence of these infections in the human race is infinitely greater than is the incidence of arthritis, which is supposed to be caused by these infections. The inescapable conclusion therefore is that in addition to an external agent of infectious, metabolic, endocrine, or even mechanical nature a constitutional susceptibility or weakness is necessary to produce the arthritis. The external agent is the more constant factor, the individual constitutional state of the patient is the unknown. When these external invaders which are operative in atrophic arthritis are mentioned, it should always be with the understanding that it is not the agent itself but only its combination with a susceptible constitution which is responsible for the disease.

A. THE ROLE OF FOCAL INFECTIONS IN THE PATHOGENESIS OF ATROPHIC ARTHRITIS

Over 100 years ago Benjamin Rush pointed out a relationship between certain infections of the body, especially the tonsils, and arthritis. He inferred that infectious material is carried from the original focus to the joints by means of the blood stream and that, therefore, an infectious focus, wherever it may be lodged, may be the source of an infection of the joint. It was argued that the great vascularity of the synovial membrane offers the most favorable site for seeding of such metastatic infections. In 1912 Frank Billings, elaborating on the problem, insisted upon a systematic search for infectious material in the body in all cases of arthritis, with the idea that by eliminating the focus a cure could be achieved.

Anatomical investigations show that the synovial membrane receives its richest supply of blood vessels at its point of attachment to the bone. The synovial membrane itself contains, in the loose areolar layer between the endothelium and the fibrous capsular wall, a great number of blood vessels and lymphatics, as well as sensory nerves. Therefore, it becomes readily engorged by vasodilatory hyperemia, especially along the reflection lines of the synovial membrane. At the present time, the role of the blood borne infection from remote foci is not considered as important as in former years. Whether it plays a part at all, and if so to what extent, can only be ascertained by a careful

analysis of a great number of cases in which such infectious foci are found in the body and are eradicated subsequently

The most common infectious foci of the upper respiratory tract are found in the tonsils, the sinuses, and the teeth Less common are foci in the genito-urinary tract What role do these foci actually play in producing the arthritis? Statistical reports regarding the frequency of established remote foci vary considerably For instance, Cecil and Archer report infected tonsils in 61 per cent and infected teeth in 33 per cent of the cases These figures would acquire meaning if it could be shown that the percentage of these infections greatly exceeds their usual occurrence in nonarthritic control cases, and that the combination of focal infection and arthritis is not merely incidental Some years ago we looked over our cases of focal infection in nonspecific arthritis, about 3,000 cases belonging to Type I and about 1,300 belonging to Type II [13] By close examination and the strictest selection, we found that removal of the foci affected the course of atrophic arthritis in not more than 15 per cent Furthermore, the improvement which followed the removal was in many cases as much attributable to the elevation of the general condition which followed the elimination of the infectious focus as to the direct effect of the focus upon the joint In the hypertrophic type, or Type II, we found that such a relation between a primary focus and the arthritis existed in 8 per cent of the cases In spite of this low incidence, it still seems advisable to include a systematic search for focal infection in the scheme of the routine examination The fact alone that the general condition may benefit greatly by removal of the focus, whether this focus is in direct connection with the arthritis or not, is good argument for adopting the policy of search and eradication

1 The upper respiratory tract

a Tonsils

Almost all tonsils show some pathological changes after the first year of life A chronic infectious condition must be assumed in a patient with enlargement of the lymph nodes and a history of repeated tonsillitis

Case C C (Female, 10 years, #41-180) had atrophic arthritis, with remission following removal of chronically infected tonsils and adenoids In this case we must assume that the chronic infection of these organs either had a direct effect upon the joints or was indirectly lowering the patient's resistance to infection

In case M H (Female, 15 years, #39 10560) it was more difficult to evaluate the effect of the removal of the tonsils and adenoids, because the treatment was supplemented by physiotherapy It may be assumed, however, that the removal of these foci was an important contributory factor in the recovery

In another case (N B , Female, 24 years, #M 12738) the improvement which followed the removal of the infectious foci was so striking that a direct connection between them and the arthritis could be assumed Tonsillectomy should be especially considered in children because of the effect of infected tonsils on the general health and upon the respiratory function

b Paranasal Sinuses

In our climate one will find the paranasal sinuses very frequently infected The incidence of such infection is out of all proportion to the incidence of arthritis The ethmoid is probably the most frequent sinus involved, but involvement of the maxillary sinus is by no means rare

c Infected Teeth as Foci of Infection

That infected teeth are a possible source of arthritis has been recognized ever since Billings (1912) laid emphasis on the concept of focal infections There is no doubt that a direct connection between infected teeth and arthritis exists in some cases We have observed a number of instances in which the removal of infected teeth brought about prompt and substantial improvement There was a time when a wholesale removal of teeth was ruthlessly practiced There is no justification for such radicalism, and one should not overlook that alveolar abscesses are frequently encountered in nonarthritic individuals Black, reporting a series of 600 normal individuals places the frequency of alveolar abscesses at 1 4 per cent In arthritis the frequency of periapical abscesses seems to be higher This is particularly true of the middle aged and older patients The question then is what kind of teeth should be suspected? The pulpless tooth without periapical translucency is less likely to be a focus of infection than the tooth which is surrounded by an abscess In general, if a tooth is manifestly infected, it should be removed

Case G C (Female) #M-9593
Age 35 years Adm April 6, 1936

This patient was admitted for painful swelling of the knees of two weeks' duration Aspiration of the knees gave a straw colored fluid The patient had a number of carious teeth, 26 infected teeth and their roots were removed When seen six months later she had no complaint and no swelling of the joints

It must be assumed in this case that removal of the teeth directly affected the condition of the joints, or that it raised the general condition to a point which made the systemic control of the infection possible

2 *The gastro intestinal tract as a source of infection in atrophic arthritis*

Dysfunction of the gastro intestinal tract seems to play a greater part in the hypertrophic than in the atrophic type of arthritis It is noted particularly that older individuals with nonspecific arthritis are unable to utilize starch In fact, it is reported that 79 per cent of the arthritic stools show excessive starch as compared with controls, and the fermentation test is positive in a high percentage of cases due to deficient starch digestion by amylase In many cases of atrophic arthritis dietetic errors may have existed for many years until the digestive deficiency finally became potent enough to produce arthritis So far as gall bladder disease is concerned, we find no higher incidence in arthritics than in other persons Operation on the gall bladder is justified only if the pathological condition of the gall bladder itself demands it Hench observed

some years ago that arthritics with gall bladder disease show considerable improvement during their periods of jaundice, and artificial jaundice produced by administration of gold salts has been used to produce a therapeutic effect

3 Ocular foci

In rare instances one will find infections of the eye and of the joints running so concurrently that some connection between the two conditions must be assumed

Uveitis and Arthritis
Case M K (Male) #40 8109
Age 30 years Adm August 25, 1946
This patient suffered from general rheumatoid arthritis for a period of six years and also had three attacks of uveitis which paralleled the acute exacerbations of the arthritis The remissions of the first two attacks corresponded to the remissions of the arthritic symptoms

We have observed 4 cases in which a connection between iridocyclitis and arthritis seemed to exist, since both conditions abated and became exacerbated simultaneously

4 The genito-urinary tract and the pelvis

At one time the connection between prostatitis and arthritis was greatly stressed by some urologists While the majority of cases of chronic prostatitis are of a nonspecific nature, it is being recognized on the whole that the prostate itself plays only a very insignificant role as the focus of infection The same may be said of pelvic sepsis in women, which is no longer considered a common producer of nonspecific arthritis

B BIOCHEMICAL DISORDERS AS SOURCES OF ARTHRITIS

1 The endocrine system

a ESTROGENIC HORMONES

A relationship between female hormones and arthritis is assumed because there are cases in which arthritis is benefited by pregnancy This is believed to be due to the effect of progestin P S Hench[6, 7] made observations on the effect of this hormone in 37 pregnancies of 22 patients with chronic articular disease Of these, 20 showed striking relief during pregnancy and for various periods thereafter In our own series, there were several cases in which an improvement of the condition during pregnancy was definitely established A certain type of arthritis occurs consistently at the time of menopause, and in some cases both muscle and joint symptoms can be controlled by estrogenic therapy Instances of this kind are reported by Bauer[2] Others are more skeptical about the connection between hormonic functions and arthritis

b THYROID HORMONE

There seems to be no constancy in the metabolic rate of arthritics However,

b Paranasal Sinuses

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3 Allergies

There is a small number of cases in which a specific allergy seems to exist to certain chemical agents or food substances which otherwise would be entirely innocuous The cause of this constitutional hypersensibility is entirely problematic

a DRUG ALLERGY

Drug allergy in general, is not uncommon, especially in older people The drugs commonly involved are the barbiturates and the coal tar products, such as aspirin and antipyrine In some cases the injection of these drugs produces arthritic irritation of anaphylactic character which promptly disappears when the drug is discontinued A 63 year old patient with a three months old non specific arthritis proved to be sensitive to aspirin, which gave him urticaria and increased the joint pain

b FOOD ALLERGY

There are also exceptional cases in which the joints react to the intake of certain foods The usual irritants are chocolate and wheat, their ingestion produces edema, exudation of the joints, and periarticular infiltration This may be accompanied by general allergic symptoms such as urticaria, asthma, vasomotor symptoms, coryza, migraine, and lacrimation Parrin (1845) was the first to point out that cases of intermittent hydrarthrosis may be due to allergy, and in 1921 Bierring[7] reported 77 cases from the literature in which the allergic element was considered, together with other endocrine factors

c ARTHRITIS AND EXUDATIVE DIATHESIS

This is hardly more than a clinical term, not a pathological entity It seems that some patients have a tendency to exudative skin reaction and other exudations Allergic urticaria and allergic dermatitis is represented by this group A relationship also exists between chronic arthritis and psoriasis In not less than 70 per cent of the patients who suffer from psoriasis, rheumatic manifestations are observed

III THE PATHOLOGY OF ATROPHIC ARTHRITIS

A GENERAL

It has been emphasized at the outset that arthritis is a systemic disease Therefore one should look for evidence of disease in all organs of the body and not in the joints alone While it is quite true that the joint manifestations are eminently in the foreground, this does not exclude that other organs and tissues may show some commensurate pathological manifestations

In its earliest stages, rheumatoid arthritis is an exudative disease, producing edema and proliferation of the connective tissues This exudative phase is followed by a granulomatous stage which is associated with the formation of

it is believed that 30 per cent of arthritics who have goiter show a relationship between hyperthyroidism and the joint condition, and that after adequate removal of the thyroid not infrequently an improvement of the arthritis follows But here again this improvement may not be the direct effect of the thyroidectomy, but may be the result of the betterment of the patient's general condition and of his increased resistance

c PARATHYROID

When Gold demonstrated the connection of the parathyroid gland with ostitis fibrosa cystica, certain American and foreign observers tried to establish a *similar connection between the function of this gland and arthritis, in some* instances the parathyroid gland was removed The results so far as we have been able to ascertain were not convincing

2 Vitamin deficiency

One should look upon vitamin deficiency as affecting the general constitution rather than as a direct causative agent It is certain that such deficiencies and chronic nonspecific arthritis are frequently found together, but it is still a question whether the vitamin deficiency is the cause or the effect of the disease

a VITAMIN C DEFICIENCY

In arthritics the blood is low in cevitamic acid, with an average of only 0 3 mg per cent as compared to the normal 1 45 mg per cent Even after adequate diet, it seems that this deficiency continues The normal requirement of vitamin C is 8 to 50 mg per day for infants, 22 to 100 mg for children, and 28 to 100 mg for adults

b VITAMIN A DEFICIENCY

This vitamin is also low in arthritics Signs of deficiency, such as photophobia, blepharitis and hyperkeratosis, are often observed in asthenic arthritics, particularly in children The normal requirement for the adult is 4,000 to 6,000 USP units, for growing children, 6,000 to 8,000 units

c VITAMIN B DEFICIENCY

This vitamin is frequently deficient in arthritics The symptoms are partly nervous and partly circulatory The urinary excretion of vitamin B which should be 30 microgram per cent may drop down to 15 microgram per cent or less in arthritics The requirement for B_1 (thiamin) should be 1 to 2 mg daily, and for vitamin B (riboflavin) 2 to 3 mg

d THE PP COMPOUND

The PP compound or the pellegra preventing vitamin of the B group may be deficient, which is manifested by gastro intestinal disturbances not infrequently seen in chronic arthritics

division of the nonspecific arthritis into two great groups, I and II, that is, the atrophic and the hypertrophic types Following Charcot, Garrot (1859) gave the first type the name "rheumatoid arthritis" and the other type the name "osteoarthritis," terms which are being used to this day

All tissues of the joint are involved in the pathological changes The synovial membrane, the capsule, the ligaments, the cartilage, the pariosteum, tendons and tendon sheaths, fasciae, aponeuroses, and even the arteries, veins, and muscles The distribution of these damages among the different structures varies greatly In general, the histological changes are the same fibrinous formations of connective tissue as are seen in other organs of the body, they are most prominent in the synovial membrane and in the periarticular connective tissue

1 The changes in the synovial membrane

The process starts in the synovial membrane, which becomes injected, thickened and hyperemic The microscopic picture of the synovial membrane shows severe damage to the endothelium, which is degenerated and transformed into flakes of fibrin, containing leukocytes and other cells (Figs 2, 3) Gradually the infected and hyperemic synovial membrane becomes thickened and

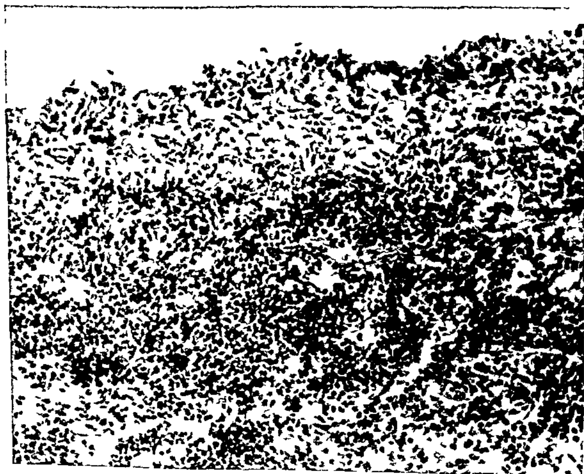


Fig 2 Proliferative arthritis synovial membrane Extreme chronic inflammatory cell infiltration Case A F

certain fibrocellular products, among these the formation of certain cellular nodes So far as the histology is concerned, these nodes seem to be identical with the Aschoff-Gepel bodies found in the heart They consist of fibrocytes in a matrix of collagen fibers and, by condensation of the fibrous bundles and the gradual loss of their cellular contents, they finally end up as a rheumatic fibrous scar To a certain extent, all organs of the body may participate in the formation of these nodes, but they do so in varying degree according to the different clinical types

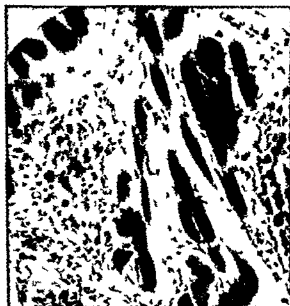


Fig 1 Rheumatic nodes in muscle (From F Klinge *Handb d Spez Pathol Anat u Histolog* Berlin O Lubarsch and F Henke 9[2] 1939)

However in all of them, the joint condition is so much in the foreground that the other organs are likely to be lost sight of Rheumatic changes have been noted in the peritonsillar tissue, in the esophagus, tongue, and lymph nodes They have also been found in the peritoneum, in the liver, in the spleen, in the kidneys, and in the muscles (Fig 1)

In the kidney the lesions appear frequently in form of a glomerulitis The enlargement of the spleen in the so called Still's disease or in Felty's syndrome is sometimes set up as a special clinical entity, but in most instances it is due to passive chronic congestion of the organ and to nonspecific proliferation of the reticulo endothelial tissues which accompanies the articular disease Visceral lesions though often only transient, sometimes leave rheumatic scars, similar to those formed in the heart or arteries Some observers even speak of a visceral type in which the rheumatic infection is confined entirely to visceral organs Not infrequently rheumatic scars in various organs are found at autopsy A H Baggenstoss and E F Rosenberg,[1] reporting on necropsies performed on 30 patients who suffered from rheumatoid arthritis, found a high incidence of rheumatic heart disease This is taken by Klinge[9] as evidence that rheumatoid arthritis and rheumatic fever are closely related, being simply different manifestations of the same disease

B THE PATHOLOGICAL ANATOMY OF ATROPHIC ARTHRITIS

For the first definite identification of the disease as a pathological entity we refer to the classical work of Nicols and Richardson [11] From the clinical point of view, however, this disease was identified as early as 1853 by Charcot, who singled out the group of so called primary progressive chronic articular rheumatism and contrasted it later with the other type now called Type II, the senile and chronic group By his description the ground work was laid for

connective tissue arises from the synovial membrane and advances from the periphery at the line of reflection toward the center of the cartilage, and eventually covers it entirely This pannus is formed through the activity of the vascular ring which encircles the insertion line of the synovia, the so called circulus articuli vasculosus of John Hunter



Fig 5 Proliferative arthritis synovial membrane Reorganization of synovial elevations presence of inflammatory cells Case A B

The pannus itself is a thin, more or less vascular membrane, sometimes a mere papillary layer with projecting tags Histologically, it consists of a very vascular granulation tissue which is infiltrated by lymphoid and plasma cells As a result of this pannus formation, the joint cavity naturally becomes reduced in size Adjacent areas of the two contacting sides of the joint constituents become matted together by numerous adhesions (Fig 6)

Simultaneously with the formation of the pannus from the synovial membrane, the bone marrow underneath the cartilage becomes transformed into fibrous granulation tissue These subchondral granulations now invade the zone of the provisory calcification They contain many chondroclasts, which gradually destroy the cartilage from below The cartilage is then caught between the synovial pannus above, and the subchondral fibrous pannus below It undergoes degeneration and finally complete destruction and disappears, so that in time the subchondral granulations meet with the granulation tissue of the pannus that comes from the synovial membrane As a result, islands of articular cartilage find themselves trapped in this double pannus formation In time, the two types of pannus, namely, the synovial and the subchondral, will fuse and then can no longer be distinguished from one another (Fig 7)



Fig 6 Joint cartilage partially destroyed Attempts at cartilaginous regeneration pannus formation Destruction of subchondral plate round cell infiltration under subchondral plate Case G S

Ultimately all the islands of cartilage disappear, this leaves the bone denuded of cartilage, and it is covered only by the proliferating connective tissue In this tissue metaplastic changes may take place The pannus may produce cartilage, sometimes it has osteogenetic properties so that bony bridges uniting one joint constituent with the other may



Fig 3 Proliferative arthritis synovial membrane Accumulation of lymphocytes swelling and separation of superficial lining, fibrin exudate Case A B

takes on a purplish color The subsynovial tissues show edema which swells and thickens the villi These tissues contain a lot of inflammatory cells, mostly proliferating lymphocytes In the subacute stage the synovial changes are essentially the same, except that we see more of the cell nodes consisting of large histiocytes, sometimes giant cells (Fig 4) The cellular rather than the fibrinoid infiltration, seen in the more acute stages, dominates the picture These granulomatous infiltrations have a central necrosis and correspond histologically to the bodies of Aschoff Gepel seen in the heart muscles (Fig 5)

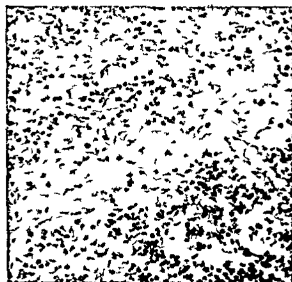


Fig 4 Proliferative arthritis the formation of a rheumatic node Synovial membrane with chronic inflammatory cell infiltration and fibrinoid swelling Case A F

The synovial exudate has a high cell count which reaches 7,000 to 10,000 per cu mm The polynuclear cells prevail in the more acute, and the monocytes and the lymphocytes in the chronic stage The synovial fluid becomes increasingly cloudy, but it never becomes purulent

2 The changes in the cartilage

Only after the changes in the synovial membrane are fully developed and have existed for some time do changes in the articular cartilage begin to appear We must look particularly to the lines of synovial reflection for the first degenerative changes of the cartilage At this stage, a synovial pannus of

destructive changes of bone follow much later, after the cartilage has been destroyed These changes appear as punched out areas of the articular surface (Fig 8) The bone marrow itself becomes fibrosed in the subchondral layers The changes in the periosteum consist merely in fibrinous infiltration and the formation of nodes One sees these nodes particularly in the neighborhood of knee and elbow

5 The changes in muscles and tendons

The outstanding feature is the marked muscle shrinkage which is due to a reflex inflammatory atrophy, indirectly, it may give rise to early static complaints in the weight bearing joints In addition to this, there is a true rheumatic myositis which is a definite clinical entity Pathologically it is essentially a proliferation of interstitial fibers, both in the internal and in the external perimysium There is early infiltration and fibrous degeneration This group also includes the so-called rheumatic myogeloses which involve the muscles secondarily to the joint involvement The nodes in the muscle are identical with the so called Aschoff Gepel bodies



Fig 9 Subcutaneous rheumatic node of the arm Case M S

The so called rheumatic myositis which appears in connection with different clinical entities of arthritis, particularly the spine, is a toxic infectious condition characterized by tenderness, induration and swelling These are mostly transitory and reversible pathological conditions Some of these clinical entities are well known, as myositis of the shoulder girdle, rheumatic lumbago, dorsal myositis, myositis of the gluteals and myositis of the neck muscles A special form is also the so called rheumatic myositis of the sternocleidomastoid or the rheumatic wryneck

6 The tendons, fasciae, and aponeuroses in atrophic arthritis

Rheumatic nodes can be seen also in the tendons and fasciae, where they appear in concrete accumulations or in diffused infiltrations A frequent location is in the posterior tibial tendon and the tendo achillis The bursae become involved in a similar pathological process The bursal sac, lined with endothelial cells, shows infiltrative changes in the subjacent tissues and often presents typical fibrous nodes with necrotic centers



FIG 7 Arthritis Fibrous and fibrocartilaginous ankylosis fusion of synovial membrane and sub chondral pannus Case A N

form, resulting in complete fusion The different types of arthritis vary greatly in this respect In some, the osteogenetic properties are greater and the bone fuses early and ex tensively In others, there is no osteogenesis at all Consequently, the end results are all kinds of adhesions and ankyloses, according to the be havior of the pannus coming from the synovial membrane and the one coming from the subchondral tissue

3 The joint fluid

Do the chemical and histological properties of the joint fluid indicate the stage of arthritis? Under normal conditions the joint fluid contains 69 per cent mononuclears, 25 per cent lymphocytes, and only 7 per cent polynuclear cells, with about 250 cells per cu mm The viscosity of the normal joint fluid is about 20 times that of water, varying within certain limits The normal synovial fluid also contains a number of ferments such as amylase, lipase, and protease

Under pathological conditions, both traumatic and inflammatory, the cell content of the joint is greatly increased Some of these cells come from the synovial membrane and others come from the circulating blood The latter also furnishes the blood cells, the plasma, the antibodies, and the fibrin One will find in acute conditions that the elements which come from the circulation prevail, and that there is a large amount of fluid with comparatively low vis cosity In chronic synovitis, however, the elements coming from the synovial membrane prevail, the fluid is more clear, has a higher degree of viscosity, and contains many synovial cells

On the whole, the blood chemistry of the arthritic joint is not materially different from the normal We find carbon dioxide, sugar, lactic acid, non protein nitrogen, chlorides, and calcium in the same proportions Kling, D H [8] found that fat in the joint fluid indicates severe injury to the synovial mem brane In nonspecific arthritis the culture of the fluid is usually negative



FIG 8 Atrophic arthritis bone changes and joint erosions

4 The pathological changes in the bone and periosteum

The only bone change which occurs in earlier stages is the bone atrophy Real

bone forming elements come mostly from the bone marrow after the cartilage has become completely exfoliated, and not from the synovial pannus

2 Still's disease

First described by Still (1897), it is a variant of rheumatoid arthritis It is a type seen in children, and is characterized by multiple joint involvement, lymphadenitis, a large spleen, and in earlier stages by fever A similar form seen in adults is called Felty's syndrome Pericarditis is often found at autopsy but without involvement of the valves of the heart itself, and there are no rheumatic nodes The synovial membrane, however, shows the same fibrinoid swelling and the same fibrinoid exudate as we see in the ordinary form of rheumatic arthritis

3 The spondylarthritis

Arthritis of the spine appears both in the atrophic and in the hypertrophic form, a detailed description will be given later At this time, we merely mention that in the atrophic type of spondylarthritis the pathological elements are identical with those we see in rheumatic arthritis in general The same rheumatoid nodes are sometimes found in the neighborhood of the spine under the skin and in the firm connective tissue of the fascia or ligaments of the back Not infrequently they are located in the neighborhood of the scapula

IV THE CLINICAL PATHOLOGY OF THE ATROPHIC ARTHRITIS

A GENERAL STATISTICS

It is claimed that in this country almost 7,000,000 people suffer from either atrophic or hypertrophic arthritis or from muscular rheumatism, and that not less than 50 per cent of these cases belong to the rheumatoid type The incidence is higher among certain races, and there seems to be a definite hereditary predisposition It seems also that climatic conditions have some effect, and that in climates which favor infections of the upper respiratory tract the incidence of arthritis is higher It is not the temperature which counts so much as the moisture The frequency of rheumatic arthritis seems to be no higher in cold climates than in warm On the other hand, dampness and low barometric conditions have a decided influence

B THE CLINICAL SYMPTOMS

1 General

Particular attention must be given to early general signs such as fatigue, lassitude, nervous irritability, and loss of appetite, which usually are noted before the first local signs make their appearance It is not enough to recognize these signs as a forerunner of the joint symptoms, it is also necessary to take them seriously enough to institute promptly certain prophylactic and supportive measures

7 The periarticular tissues in atrophic arthritis

a Subcutaneous Nodes

The most favored location of the rheumatic nodes however, is the subcutaneous tissue, particularly over the muscle fasciae (Fig , 9, rheumatismus nodosus) They consist of connective tissue with chronic inflammatory cells and have a tendency to fibrinoid degeneration The nodes have an acute, exudative and a chronic, granulomatous stage Some of them may disappear without any trace, others terminate in permanent scar formation which sometimes contains calcium deposits. Coburn[4] makes the statement that these subcutaneous nodes can be found in as many as 15 per cent of the cases Sometimes they appear very early in the disease, and sometimes quite suddenly, at the elbow, hand, knee, hip, or shoulder blade, and are often symmetrical They are tender to pressure, but show no spontaneous pain

b Changes in the Capsular Apparatus

Synchronous with the changes in the synovial membrane, the capsular apparatus shows proliferation of its connective tissue layers All changes blend indistinguishably with those of the synovial membrane Some of the accumulations of fibrous tissue can be considered as diffuse rheumatic nodes They result in a thickening and infiltration of the reinforcing ligamentous apparatus of the joint

8 The peripheral nerves in chronic atrophic arthritis

Recently Freund, Steiner, Leichtentritt and Price,[5] examining the peripheral nerves in 5 cases of arthritis, found in three of these small nodules of characteristic microscopic structure The nodules were located in the perineurium and were 0 1 to 0 3 cm long They showed a central acellular zone of homogenous coagulated material and a peripheral outer zone which consisted of numerous lymphocytes and plasma cells, similar to those in the subcutaneous tissues and elsewhere

C THE SPECIFIC PATHOLOGICAL TYPES OF ATROPHIC ARTHRITIS

1 The ankylosing type

A specific ankylosing type of arthritis, or arthritis ankylopoetica, is a clinical entity It involves the peripheral joints as well as the spinal column, and is characterized by a more rapid degeneration of the cartilage following the pannus formation The granulations soon fill the entire joint space and form a fibrous ankylosis, but the degeneration and disappearance of the cartilage must precede this occurrence The destruction of cartilage is rapid, and the fibrous tissue formed by the pannus is especially firm It is in this type of rapid joint destruction that metaplastic bone formation and bony ankylosis are seen The

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The sedimentation time Short, Vienes and Bauer[1] found that in the acute stage the sedimentation time is accelerated in 90 per cent of the cases A left shift of the Schilling test was found in 87 per cent This acceleration is of diagnostic value for the differentiation from noninflammatory conditions As the case enters the more chronic stage, the sedimentation time becomes normal, and the Schilling test no longer shows a left shift Rapid increase in sedimentation is an unfavorable prognostic sign

2 Early local symptoms

a SUBJECTIVE SIGNS

The first subjective signs are transitory The patient complains of paresthesias, numbness, and tingling before any local joint manifestations appear These complaints depend a great deal on the weather, particularly on the changes in barometric pressure, the patient becomes sensitive to dampness and rain, which he is frequently able to predict on the basis of his subjective complaints It is very significant that the subjective signs are worse in the morning on arising, and a common statement of the patient is that the joints limber up after he has moved around for some time On the other hand, exertion and strain have a detrimental influence, while rest relieves the condition, thus the patient after having been more comfortable in the morning again becomes uncomfortable as the day progresses He does not seem to tolerate one position for any length of time He has the urge to change position and to move about

The joint pain varies, it follows the spread of the disease from joint to joint, but in degree it does not parallel the objective changes It is characteristic for this type of arthritis that involvement of the joint is usually symmetrical, and that it proceeds from the more peripheral to the more proximal articulations The pain is by no means constant in intensity It fluctuates with the time of day and the season of the year

b THE EARLY OBJECTIVE SIGNS

1) Swelling and tenderness The first objective sign is the spindle shaped swelling of the proximal interphalangeal joints of the fingers (Figs 10, 11)



FIG 10 Early objective changes swelling of interphalangeal joints
Case E W

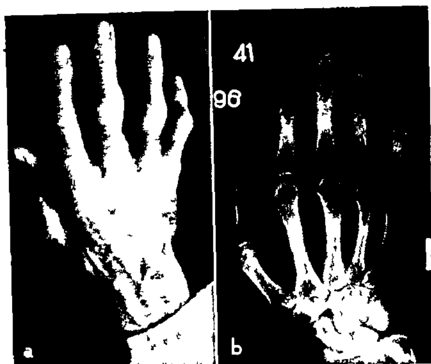


FIG 11 Atrophic arthritis Case H W (male), #41 5295, 49 years April 1941 Complaint of pain and stiffness of the ankles for one year followed by stiffness and pain in finger Swelling of midphalangeal points X ray narrowing of midphalangeal joint space a) Finger joints b) X ray Note joints of fifth finger

They are tender to pressure, they may feel hot, but there is no redness The extremity as a whole feels rather cold and clammy, due to vasoconstriction The involvement of the large joints follows, in line of frequency they are the knees, ankles, wrists, and shoulders In the meantime, the sensitiveness to weather and barometric changes increases and the general condition deteriorates, there is loss of weight and appetite and loss of sleep as the patient is kept awake by pain and discomfort

2) Muscle atrophy Muscle atrophy parallels the development of joint swelling It progresses rapidly and therefore makes the joint stand out more plainly so that it seems to be unduly enlarged The muscles of the larger joints at first maintain the articulation in midposition by spastic reflex contracture, which is the principal cause for the subsequent deformity The atrophy is inflammatory and not an atrophy of disuse, and the muscles maintain an increased spastic tone Recognition of the spastic nature of the contracture in the beginning phases of arthritis is of the greatest importance for the correction of the contracture

3) Contractures *The contractures of the fingers and wrist* In general, two types of arthritic contractures of the fingers may be distinguished The *clawhand contracture,* and the *pill roller hand contracture*

In the clawhand contracture the metacarpophalangeal joints are hyperextended, and the interphalangeal joints are flexed, similar to what one sees in paralysis of the median and ulnar nerves (Fig 12)

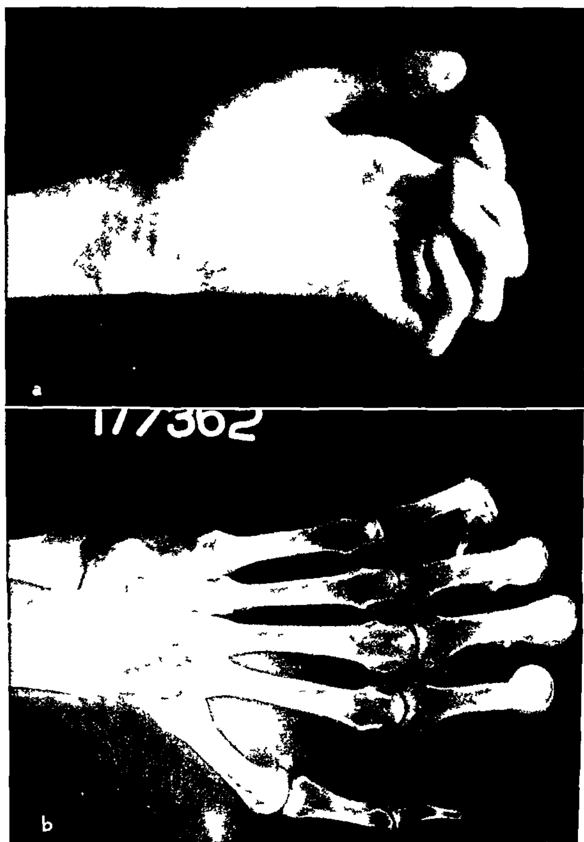


Fig 1? Atrophic arthritis clawhand contracture Case E L (female) #43 1?991 29 years December 1943 Rheumatoid arthritis involving left ankle wrist and fingers duration six month Wrist ankylosed metacarpophalangeal joints extended thenar atrophy a) Contracture b) X ray showing ankylosis of wrist

The pill roller hand deformity consists in flexion of the metacarpophalangeal joints and extension or hyperextension of the interphalangeal joints The latter stiffen rapidly in extension, due to early capsular shrinkage or fibrous ankylosis (Fig 13)

Contractures of the lower extremities In the lower extremities swelling and pain are particularly noticeable in the ankles and knees (Fig 14) The degree of contracture is not commensurate with the degree of swelling Frequently we see a large joint with comparatively free movement, whereas joints with little or no enlargement may be in a state of severe contracture This is particularly true in the case of multiple hydrops of the larger articulations where the swelling becomes especially marked



Fig 13 Atrophic arthritis pill roller hand contracture Case C E (female) #H 9952 34 years October, 1933 Early arthritis seven weeks duration Extension deformity right wrist pill roller hand deformity right flexion contracture of metacarpophalangeal joints



Fig 14 Atrophic arthritis Flexion contracture of knees swelling of ankles

3 The later signs

It is typical of the nonspecific atrophic arthritis to run a course of *exacerbations and remissions* These remissions concern especially the subjective symptoms of pain and stiffness, and they are difficult to account for The objective signs of thickening and infiltration are more persistent

In general, one will find that the arthritic changes develop to a certain degree and then become stationary, the subjective symptoms subside or "burn out" This is so irregular and variable in different cases, however, that a prediction when spontaneous relief from subjective symptoms will occur can seldom be made As a rule, by the time these symptoms have disappeared or "burned out," there is already a considerable irreversible damage in the joint

a THE DEFINITE CONTRACTURES AND DEFORMITIES

In the early stages, the arthritic contractures are due to reflex spasm Purely spastic contractures are amenable to correction by mechanical means As the disease progresses, however, the contractures become more resistant and less amenable to mechanical correction This is due, first, to the changes which occur in the joint itself and in the capsular apparatus and, secondly, to adaptive changes in the muscles as they accommodate themselves to their shortened position Consequently, during the development of the deformity the joint position is dominated principally by the muscle spasm, and correction is still possible In the later stage, when capsular shrinkage has taken place and intra articular destruction of bone and cartilage has occurred, the situation is very different Then, only operative measures will be able to change the position of the joint



FIG 15 Atrophic arthritis Extreme and intractable late contractures of fingers and hands Case J N

There are types of arthritis in which the joint cartilage melts away more rapidly, and fibrous and often bony ankylosis results This occurs not only in the large joints but also, and too often, in the small joints of the fingers, particularly in the pill roller type of contracture Only energetic and timely measures during the initial period preceding actual joint destruction save such hands or extremities from falling into utter disuse (Fig 15)

4 Unusual symptoms of atrophic arthritis

a SUBCUTANEOUS NODES

Their frequency is placed by some as high as 20 per cent Usually they are not tender On the whole, they indicate a rather poor prognosis Localization in the skin is most common, less often they are found around the joint or in the joint capsule itself (Fig 16)



FIG 16 Atrophic arthritis Clawhand contracture subcutaneous nodes Case J R

b Skin Changes

One finds the skin usually clammy, and sometimes it takes on a bronze hue Many of the patients have a certain exudative diathesis, psoriasis is not an infrequent complication

c Visceral Changes

While they do not cause definite clinical symptoms, the gastro intestinal tract is often sluggish and shows poor carbohydrate and fat absorption A frequent result of these digestive disorders is visceroptosis The patient suffers from constipation and intestinal stasis, if the diet is abundant in starch, he may have considerable digestive difficulty The colon should receive a good deal of attention, as it is usually atonic Today we no longer accept resection of the colon as was introduced by Arbuthnot Lane many years ago, but we find that the incidence of gastro intestinal disturbance is much higher in atrophic arthritis than in other diseases, and evacuation of the colon is one part of the treatment

V THE X RAY FINDINGS IN ATROPHIC ARTHRITIS

Because x ray evidence appears comparatively late, it cannot be relied on for the early diagnosis of arthritis, but it is a good indicator of the progress of the disease

A THE EARLIEST X-RAY SIGN

Atrophy of the bone is the earliest x ray sign (Fig 17) Later, the fusiform swelling of the joint can be made out as the disease progresses Still later, the





Fig 18 Atrophic arthritis joint erosion Case W H

←

Fig 17 Atrophic arthritis Marked regional atrophy narrowing of interphalangeal joint spaces

narrowing of the joint space can be seen, together with the mottled appearance of the atrophic joint constituents

B LATER SIGNS

When the cartilage succumbs to the destructive effect of the pannus, we see irregularity of the outline, punched out places and erosions (Fig 18)



Fig 19 Atrophic arthritis Complete destruction of the interphalangeal joint of the index finger partial destruction of the metacarpophalangeal joint of the middle finger (Courtesy A I Friedman Green Bay Wisconsin)



Fig 20 Atrophic arthritis of the left hip with fibrous ankylosis Case V W (female) #38 19666 30 years September 1936 Atrophic arthritis left hip with fibrous ankylosis X ray narrowed joint space with irregular outlines Treated later by arthroplasty



Fig 21 Atrophic arthritis of the knee bony ankylosis Case J L a) Anteroposterior b) Lateral

Finally the joint becomes entirely destroyed It is filled with fibrous tissue, and complete obliteration of the joint may result (Fig 19 courtesy, Dr A I Friedman, Green Bay, Wisconsin) Sometimes the process of destruction does not stop until the joint is completely obliterated and all vestiges of cartilage have disappeared (Fig 20) The osteogenetic activity of the bone marrow then may produce complete bony ankylosis, very much as we see in the pyogenic

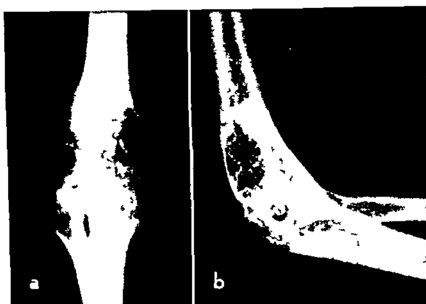


FIG 22 Ankylosing arthritis of the elbow Case J L
a) Anteroposterior b) Lateral

joint The obliteration may be so complete that the bone trabeculae flow in continuous trajectories from one bone to the other, and become reoriented on static lines (Figs 21 and 22)

VI THE DIFFERENTIAL DIAGNOSIS

A IN THE ACUTE STAGE THE FOLLOWING CONDITIONS MUST BE DIFFERENTIATED

1 Rheumatic fever

Here the decision lies with the history, the involvement of the heart, and the response to salicylates

2 Gonorrheal arthritis

There is a history of gonorrheal infection It is usually monarticular, the joint is extremely painful and swells rapidly Also it prefers certain articulations such as the knee, wrist, and metatarsophalangeal joints

3 Traumatic arthritis

There is a history of trauma, and the symptoms develop gradually and remain constant The location is monarticular

4 *Septic arthritis*

The course is stormy A general septicemic condition may develop The joint destruction is much more rapid, and the x ray picture shows changes comparatively early

5 *Tuberculous arthritis*

It is also monarticular and of slow development There is a positive tuberculin test, and the x ray changes have some characteristic features

6 *Hemophilia*

A hemophiliac gives a history of bleeding and of hereditary incidence Usually only one joint is involved There is rapid swelling which is very painful and which follows immediately after injury Fever is absent The x ray picture shows punched out areas of the margin which are very suggestive

7 *Gout*

Gout occurs more frequently in people 40 to 60 years of age, whereas the rheumatoid arthritis is a disease of younger people However, the differentiation is not always easy, particularly in the acute form Gout often occurs in several joints Suggestive are the rather rapid development of redness and swelling which occur periodically, the intense pain, the x ray changes and, finally, the high uric acid content of the blood

B IN THE CHRONIC STAGE OF RHEUMATOID ARTHRITIS THE FOLLOWING CONDITIONS MUST BE DIFFERENTIATED

1 *Senescent, degenerative or hypertrophic arthritis*

It involves higher age groups, is more often monarticular, and shows no actual obliteration of the joint Differentiation between rheumatoid and hypertrophic types is complicated by the fact that mixed types are not infrequent These are seen in patients of middle age or older, in whom an atrophic arthritis becomes superimposed upon a pre existing hypertrophic type

2 *Tuberculosis*

This is monarticular, it develops slowly, and shows early joint destruction which is more extensive than is the rule in the atrophic arthritis J G Kuhns[10] reports only 14 cases out of a total of 2,268 arthritic patients diagnosed as monarticular atrophic arthritis Of these 14, only five proved to be atrophic arthritis, the others finally proved to be tuberculosis

3 *Chronic gonorrheal arthritis*

Unless there is a definite history of primary infection, this condition is often difficult to distinguish from chronic rheumatoid arthritis Usually the joint destruction in gonorrheal arthritis is more rapid and more extensive

The monarticular type is much more frequent than the pluriarticular type Its preference for certain large joints, in contrast to involvement of the small joints of the fingers one sees in the atrophic arthritis, is also a help in the differential diagnosis

REFERENCES

1 Baggenstoss A H and Rosenberg E F *Am J Path* *35* 503 1943
2 Bauer W *New England J Med* *221* 533 1939
3 Bierring W L Quoted by Berger H, *J A M A* *112* 2402 1939
4 Coburn A F *The Factor of Infection in the Rheumatic State*, Baltimore, Williams & Wilkins Co 1931
5 Freund H A Steiner B, Leichtentritt B, and Price A E *Am J Path*, *18* 865 1942
6 Hench P S *Canad M A J* 2 394 1938
7 ——— Acute and Chronic Arthritis *Nelson s Looseleaf Surgery*, 1925
8 Kling D H *Am J Surg* *6* 71 1929
9 Klinge F *Virchows Arch f path Anat* *278* 438 1930 *281* 701 1931, *286* 344, 1932
10 Kuhns J G *New England J Med* *232* 128 1945
11 Nichols E H and Richardson F L *J Med Res*, *21* 149 1909
12 Short C L Vienes L and Bauer W *J A M A*, *108* 2087 1937
13 Steindler A *Proc Am A Study Rheumat Dis* First Meeting Cleveland, June 11 1934

Lecture II

ON THE GENERAL TREATMENT OF ATROPHIC ARTHRITIS

IN THE description of the clinical symptoms particular stress was laid upon the general premonitory signs of lassitude, fatigue, loss of weight, and certain early local signs such as coldness, paresthesia, and stiffness of joints It is regrettable that too frequently neither the patient, nor even the physician, takes these signs seriously enough to institute prompt measures Yet, almost the only hope for recovery rests with the early recognition and evaluation of these symptoms Such is the pace of progress in many cases that irreversible changes have become established before one is aware of the seriousness of the condition

The old truism that the first consideration should be given to the patient, and the second to the disease assumes special significance in the case of atrophic arthritis By and large, too little attention is paid to the needs of the patient's constitution, which after all shoulders the burden of defense against the invading organism or toxic agency The first objective therefore is to raise the patient's state of health to a point of natural resistance able to convey an adequate degree of immunity against the spread of the disease, the next objective is the care of the disease itself and of the affected joint, and then the care of secondary deformities and disabilities A third objective too often neglected is the social rehabilitation Following the physical reconstruction of the deformities or disabilities acquired through the disease, there is still much to be done in rehabilitating the patient by occupational therapy and vocational training

I THE HYGIENIC TREATMENT

A REST

Complete mental and physical rest is the first thing to provide for the patient It means long hours of sleep, rest periods during the day, protection against worries anxiety, responsibilities and excitement In other words, the patient should be treated as one would treat a cardiac case and, above all, should not be given an unnecessarily gloomy prognosis He should know that at least 70 per cent of the cases of this type can be greatly benefited if not cured by some means or other

To provide this rest and comfort medicinal sedation is required In milder cases aspirin or phenacetin will suffice Many patients, however, require more energetic sedation during the acute period Restless or apprehensive patients should be given barbiturates preferably one half grain of phenobarbitol two or three times a day For local pain, codeine sulfate may be administered in

doses of one quarter to one half grain several times a day It is, however, much better to depend for the control of local pain on physiotherapeutic measures rather than on sedatives

B DIET

It was formerly believed that the diet should be drastically restricted (Pemberton), particularly in regard to carbohydrates The general opinion now is that restriction of the diet is not necessary, and that animal proteins should not be withheld, particularly in undernourished and anemic patients The diet should be rich not only in vitamins and mineral content but also in protein On the other hand, there is some reason for restricting the carbohydrate intake If the patient is recumbent, he cannot take care of carbohydrates since his muscular activity is restricted The caloric intake of the patient should be 2,000 to 3,000 calories He should have at least one quart of milk, whole wheat or rye bread, butter, fresh fruit, green vegetables, as well as an abundant amount of proteins

Vitamins must be added to the diet because the arthritic is lacking in vitamins B and C, and, often, in vitamin D The vitamin D deficiency of arthritics has been known for many years Marshall G Hall, C O Bayles and Soutter showed that 65 per cent of the arthritics are vitamin D deficient, and that the deficiency parallels the severity of the case

So far as vitamin C deficiency is concerned, it was found by Rhinehart* to exist in 75 per cent of the cases, the blood level being below 0 5 mg per cent This author recommends adding 100 mg of vitamin C to the general diet There is no reason why arthritics should refrain from fresh fruit, fruits and vegetables are beneficial because they furnish vitamin C which the patient is lacking Vitamins C and B can be given by intravenous administration, the latter as B complex in the amount of 1 cc of a 5 cc ampule once a day, the course of treatment being 25 cc Vitamins A and D may be given by mouth

C APPETITE AND ELIMINATION

The anorexia can be taken care of by administration of iron, quinine and strichnine, and even small doses of insulin For habitual constipation colonic flushing should be applied temporarily and should be followed by a mild cathartic of saline nature A regular habit of elimination should be developed We would warn against the continuous use of mineral oil because of the high solubility in oil of vitamin A, which may easily be carried away by it and thereby made unavailable for the body

D BLOOD TRANSFUSION

For building up the general resistance, there is no better means than fractional transfusions of blood, such as we use for the same purpose in tuberculosis and osteomyelitis As many of these cases show secondary anemia, transfusions of 200 to 300 cc given once or twice weekly are particularly appropriate

E POSTURE AND BODY MECHANICS

Many ambulatory patients show poor posture and generally faulty body mechanics, mainly due to muscular weakness The respiratory capacity is greatly reduced There is digestive sluggishness and constipation Therefore, whenever possible, the treatment must include corrective postural exercises in or out of bed, consisting of systematic training of chest muscles and respiratory muscles as well as the muscles of the abdominal wall

II GENERAL BIOLOGICAL MEASURES

A THE REMOVAL OF FOCI OF INFECTION

We mention this under general treatment because the removal of the foci has its principal effect on the general condition of the patient We do not believe that a focal infection directly influences the joints, except in a small minority of cases, or that one affected joint has a similar effect upon the other joints of the body The removal of a focus must be considered rather as a general measure which elevates the patient's resistance

In former years, the opinion was entertained that these remote infectious foci exercise a dominating influence on the disease Consequently they were removed radically and often very indiscriminately Obviously, only those foci should be removed in which an active infection is definitely established The safest policy is to remove a focus only if removal is indicated by the local condition itself, to prevent absorption of toxic material, irrespective of the existing arthritis, this should apply, for instance, in the case of an abscessed tooth, or an empyema of the nasal sinuses If several foci should be found, as is frequently true the question arises, which should be removed first The sequence depends on the degree of infection in each focus, other factors being equal, it would be Tonsils, sinuses, and teeth

1 Tonsils

Only if they are definitely infected and if their infection results in repeated attacks of tonsillitis should their removal be advised The removal should be clean surgically, and no stumps should be left We believe that age is no contraindication, but one should warn against removal of tonsils in children in the fall or late summer especially if there is an epidemic of poliomyelitis or influenza

2 Paranasal sinuses

Suspected sinuses should be trans illuminated and x rays taken If pus is present, especially in the maxillary and frontal sinuses, it should be drained The ethmoid sinus is most frequently involved, and the suction treatment of Proetz seems to be very successful It is the general attitude of the otolaryngologist to be rather conservative, except where there is a definite empyema otherwise drainage can be accomplished by shrinking the nasal mucosa with ephedrine or other drugs One should not neglect to inspect the posterior

pharyngeal wall for roughness and moisture, and particularly for a postnasal drip In cases of polyps or thickened mucosa or sinus headaches with purulent postnasal discharge a good deal of improvement of the general condition can be obtained by proper treatment of the sinuses

3 Teeth

The former practice of extracting teeth en masse has been given up and, largely upon the insistence of the dentists themselves, a more conservative attitude is now observed We do not believe that the teeth should be extracted during the acute phase or during an acute exacerbation of the arthritis, and not more than one or two infected teeth should be removed at one sitting A pulpless tooth with absorption of the root, or one which shows a rarefying osteitis, indicating infection, should be removed

4 Gall bladder and prostate

They are so rarely the cause of arthritis that we may safely say that the question of treating them surgically must be decided entirely on the merits of the local condition

B TREATMENT BY VACCINE

Autogenous and stock vaccines became immensely popular as the idea of focal infection took hold Autogenous vaccines are prepared from an infected focus of the patient, while the stock vaccine contains a conglomeration of all kinds of streptococcus and other strains A new impetus was given the vaccine treatment by the method of Crowe[9] This investigator devised an extremely elaborate technique of application It was based principally upon the idea that reactions to the vaccine injection, general or local, should be avoided and that, consequently, the doses should be kept under the reaction level This was quite in contrast to the former technique in which reactions, both local and general, were expected

Crowe's vaccine consists of polyvalent stock vaccine containing 155 types of streptococcus and staphylococcus and Micrococcus deformans The initial dose was 100,000 cocci, the principle of the treatment was to avoid consistently any reaction In 1940, Crowe reported on 7,000 cases treated with his vaccine, 67 per cent of which had some type of arthritis

He found, all in all, that 50 per cent of the cases showed excellent results, and 30 per cent were improved He was careful to point out that the patients become sensitized so that, as a rule, they require smaller and smaller doses Starting with 100,000 streptococcus and staphylococcus bacteria, he cuts the dose down until it becomes as low as 500 bacteria per dose given at regular intervals It seems that much better results were obtained by Crowe's method than by any other administration

Still, it is difficult to evaluate the treatment For instance, Sidel and Abrams,[23] studying the effect of vaccines, used normal saline for control The controls received daily subcutaneous injections of 0 5 cc of normal saline

solution, and were observed for a period of six months to four years In a series of 33 control cases of rheumatoid arthritis, improvement was claimed in not less than 72 per cent

III THE GENERAL MEDICINAL TREATMENT OF ATROPHIC ARTHRITIS

We have stated that there is no specific drug for arthritis, at least not in the same sense as the salicylates for rheumatic fever or quinine for malaria There is a formidable array of drugs which at one time or another have been used and have been credited with therapeutic value Most of these drugs have gone into discard one by one and the select few which have remained still cannot claim any degree of specificity

One may divide these drugs into three groups One group acts upon the vasomotor system as a vasodilator and increases the hyperemia This results in recession of such reversible changes as are still present in the affected joint The second group is purely analgetic, such as the salicylates, their effect is too temporary to be mentioned except in passing The third and largest group is one which stirs up and excites the reticulo endothelial system, stimulating it into increased production of antibodies in the tissues These latter drugs may be foreign proteins or metals or any other substance affecting the reticulo endothelial system of the body

A THE VASODILATOR GROUP

Mecholyl and histamine

Vasodilation overcomes peripheral coldness and clamminess of the extremities The vasodilatory effect of *histamine* on the peripheral vessels remains in force for one or two hours and produces at the same time a marked analgesia, it is particularly useful in painful joints or in fibrositis of muscles When applied by iontophoresis, the indifferent large electrode is usually placed on the back, and the active or positive electrode, consisting of copper wire screened in gauze pads 2 x 2 inches in size is applied to the limb

The *choline* substance stimulates the parasympathetic nerves which are vasodilators Mecholyl is used in 0 5 per cent solution in a current 25 to 30 milliamperes for 20 to 30 minutes Combinations of mecholyl and histamine are also recommended In a review of the literature, Kling[18] cites 376 cases with 343 favorable results

B THE STIMULANTS OF THE RETICULO ENDOTHELIAL SYSTEM

1 Foreign proteins

Typhoid vaccine is still extensively used One starts with 25 to 200 million dead bacilli as the initial dose A reaction in form of chills and rise in

temperature is expected Some reports state improvement in as high as 20 per cent From our own experience, we believe the improvement is mostly subjective and certainly always temporary, and we question very much the value of this method The treatment with foreign proteins of other nature such as casein practically tells the same story Reactions occur in the form of rise in temperature or chills, the beneficial effects soon subside, and the condition remains unchanged

2 Bee's venom

This is also a foreign protein The only reason we mention it is because it has been known among the laity that keepers of bees seldom suffer from arthritis Bee's venom is a dilator of the capillaries and thus is similar to histamine It is still given occasionally in rheumatiod arthritis in doses of 0 1 cc in solution, twice a week, for a total of 25 injections Cases of improvement have actually been observed Of 100 cases reported by Kroner *et al*[19] marked improvement was claimed in 35 per cent and moderate improvement, in 38 per cent

3 Minerals

Iodide

Some years ago, iodides were very much in use, they are seldom applied now The drug acts as an irritant to the reticulo endothelial system The most popular is the amydoxyl, a drug now practically abandoned In 1934, Wheeldon [7] claimed improvement in as high as 98 per cent of his 46 cases

Sulfur Drugs

These also had some ardent followers for a time, their use was based upon the idea that there is a deficiency of sulfur in arthritis, and that the patients show a low cystine content However, this concept has now been entirely refuted For those still interested in the administration of these drugs it may be mentioned that they can be given in doses of 1 to 2 mg , both intravenously and intramuscularly, this may gradually be increased to 15 mg per dose The injections are given two to three times per week until about 300 mg are reached in the adult To us, the effect of these drugs seems doubtful, in spite of the fact that favorable results were reported from time to time In our clinic a study was made of 37 cases of both the hypertrophic and atrophic types The drugs used were diasperol or sulisocol, according to whether application was intravenous or intramuscular Of 23 cases reported, 13 showed apparently good or fair results so that it appeared, at least at the moment, that the sulfur had some beneficial effect However, the effect was so temporary and the remissions caused by it resembled so much natural remissions of the disease that we still doubt the efficacy of these drugs

Phosphates

Acid potassium phosphate has been recommended lately by Crowe[10] for use in arthritis It is used on the theoretical grounds that it acts as a buffer in chronic synovitis where the fluid is alkaline, the pH here being above 7 4 In contrast to this, the acute arthritic joint has acid fluid Crowe aspirates the joint and introduces acid potassium phosphate in 1 per cent solution, 20 cc for the hip and for the knee Reports on 284 joints so treated state that 68, or 24 per cent, became painless with full motion, 102, or 36 per cent, had objective signs of relief with decrease of swelling and improvement, 114, or 40 per cent, showed a temporary subjective relief

Gold Therapy

The use of gold in the treatment of arthritis was introduced by Forestier,[1] there is probably no other single drug used in the treatment of arthritis today which has gained such prominence as have the gold salts They are the most potent irritatives and stimulants of the reticulo endothelial system employed today

Selection of the Gold Preparation

All gold preparations contain a high proportion of gold, 40 to 50 per cent, combined with a sulfur radical Some of these are soluble in water, and some are insoluble and are used in colloidal suspensions We prefer the thiomalate and thiosulfate of gold because they are soluble drugs, they are also known under the names of myochrysine and solganol

Administration of the Drug

The drug is administered in small doses, ranging anywhere from 10 to 15 mg or even less, given in frequent intervals, until a total of 1000 mg is reached The high toxicity of the gold, however, requires a strict and careful program We follow the plan of Comroe[3] which is as follows The first four injections are given in intervals of three or four days, 5 mg each All subsequent injections are given once a week, in doses increasing from 10 to 50 mg, according to the patient's response If improvement is definitely noted after the first course of gold which lasts about three or four months, then the patient is given a rest period of four to eight weeks, if a relapse occurs, the gold therapy is resumed The amount of gold injected should vary between *850 and 1000 mg, which is equivalent to 425 to 500 mg of metallic gold* If the soluble thiosulfate is used, the dose can be one third larger It is important to proceed cautiously with the increase of dosage to avoid toxic reactions

Toxic Reactions to Gold

Some years ago the mortality from the treatment was as high as 3 per cent It has now been lowered to less than 0 5 per cent, but the drug is still dangerous,

and in slightly less than 20 per cent of the cases it must be discontinued The metal is deposited in the liver, spleen, kidneys and skin A small amount of gold is excreted through the intestine and bronchial mucosa, but most of the gold is excreted by the kidneys When large doses of gold are given, such as 100 mg, approximately 20 per cent of the gold is excreted per week, about 80 per cent of the injected gold being retained in the body, which means that gold may be found in the urine and blood plasma for many months, in gradually diminishing amounts The most common toxic manifestations are pruritis, dermatitis, stomatitis, and gastro intestinal symptoms More violent toxic reactions are fever, dizziness, urticaria, and pleurisy Some patients are allergic and hypersensitive and must be treated with special care The treatment must always be controlled by constant and adequate laboratory examination of the blood, the sedimentation rate, the urine, and the renal function Occasionally there appears an agranulocytosis and purpura, which is a real danger, a complication which Forestier observed in seven out of 500 cases Because of its toxicity, gold should not be used as routine treatment for arthritis but only after other methods have failed

Precautions

The patient should be questioned for any signs of toxicity, rash, itching, sore mouth, metallic taste, jaundice, or loss of appetite Blood counts and urinalysis must be carried out before injection A history of purpura or agranulocytosis is a contra indication, so is also renal or hepatic disease, pregnancy, hemophilia, severe diabetes, colitis, severe anemia, as well as eczema and chronic dermatitis

The Effect of Gold Salt Therapy

If rheumatoid arthritis is treated at an early stage, when the inflammatory process is limited to the soft tissues, then the improvement after administration of gold salts may be very striking It is not nearly so spectacular in advanced cases in which there are already considerable joint changes The first signs are relief of joint pain, softening and gradual disappearance of swelling, and increase of joint mobility There is a marked improvement in the patient's general condition Relapses following gold therapy are seen most often within six to 12 weeks following discontinuation of the drug

Statistics on Gold Therapy

According to Comroe,[8] gold treatment is probably the most valuable single drug treatment in rheumatoid arthritis Forestier[12] reported 500 cases, of which 70 to 80 per cent showed definite improvement Fifty per cent of the early, and 20 per cent of the late cases of two or more years' duration were considered as cured Statistics on a large number of cases are those of Hartfall and Garland,[14] who report on 900 cases, of which 9 9 per cent were cured and 56 8 per cent were markedly improved Gardner[13] reports 250 cases, of which

70 to 80 per cent responded favorably to treatment Price and Leichtentritt[1] found even a higher percentage of favorable results In the milder cases 93 per cent improvement with disappearance of all symptoms, especially pain and stiffness, in the moderate cases 80 per cent improvement, and in the severe cases 40 per cent In 1945, Cohen and Goldman[7] reported 259 cases, with marked improvement in 48 per cent Less favorable is the experience of Browning, Rice, *et al*,[3] who cite 47 cases observed in their six years of experience with gold therapy They found that only 23 per cent continued in an improved state after treatment, 62 per cent showed no appreciable changes, and 15 per cent became worse They also found a high incidence of toxic reactions —as high as 52 per cent Most of these were of minor nature, however although there were two cases of exfoliating dermatitis and one death Our experience with this drug although covering only a small number of cases, has on the whole been favorable The majority of cases show an improvement, consisting in the lessening of pain, more freedom of motion and, especially, a greatly improved general condition and feeling of well being

4 Antireticulo cytotoxic serum

Several years ago Bogomoletz of Russia prepared a filtrate from the reticulo-endothelial system recovered from bone marrow, the spleen, and other places, which he believed contained the antitoxic substances of defense against infection This serum has recently been used for the treatment of atrophic arthritis, some observers reporting favorable results[11] Up to the present time there are, however, no statistics on hand on definite end results from the use of this drug

5 Penicillin and the sulfonamides

The general opinion is that neither penicillin nor the sulfonamides have any effect on arthritis, although occasionally satisfactory results from intra articular use are being reported

6 Medicinal use of vitamins

In addition to the use of vitamins as part of the diet, some of them have been used in large doses in the medicinal treatment of arthritis Abrams and Bauer[1] gave vitamins in massive doses to patients in stages when the pathological process was still reversible They administered from 50,000 to 1,000,000 USP units of vitamin D and came to the conclusion that these massive doses produced subjective improvement in a number of cases, but they were not able to obtain any objective results In the same category belong ergosterol and ertron Activated ergosterol in doses of 100,000 to 600 000 units was used by Snyder and Squires[5] They found no serious toxic manifestations and observed some improvement in the systemic conditions of the patient consisting in increase of weight and some decrease in the local swelling Ertron, which is activated ergosterol has of late received a good deal of public notice,

and sweeping claims have been made regarding the effect of this drug However, a late report of the American Medical Association expressed the opinion that these claims were not justified

Recently, also, in a study of 180 cases, Magnussen, McElvinny and Logan[20] introduced a new drug, the steroid complex, which is a combination of activated and vaporized ergosterol administered in capsules of 5 mg, which equals 50,000 USP units of vitamin D The maximal daily dose in most cases was 30 mg, equivalent to 300,000 USP units of vitamin D They believe that this remedy has a palliative effect in many cases of rheumatoid arthritis, and report good results in 32 3 per cent of the cases On the other hand, the report of Slocumb,[1] covering a study of 14 patients treated by viosterol in dosages of 50,000 to 380,000 units, showed that the relief was only partial, and exacerbations occurred even while the patient was still taking the vitamin He concluded that so far as the lasting improvement is concerned the drug was useless Besides, there is the danger of hypercalcemia, in their series, Abrams and Bauer[1] cite a considerable number of cases who developed calcinosis

7 The pituitary and adrenal hormones

In April, 1949, Hench, Kendall, Slocumb and Polley[15] announced the use of an adrenal hormone which was called Compound E and later Cortisone, and of a pituitary hormone (ACTH or adrenocorticotropic hormone) in the treatment of rheumatoid arthritis

Cortisone had been previously isolated by Kendall from the adrenal glands This Compound E or Cortisone had been administered to 16 patients which were severely or moderately affected and in each case a startling improvement was observed Similar clinical results accompanied by various biochemical effects were obtained by the pituitary hormone, ACTH Obviously the latter was functioning by stimulating the production of the adrenal cortical hormone

It is believed that the action of the hormone on hyaluronidase affects the cell permeability and that the hyperadrenal state suppresses responses of the mesenchymal tissue and so interferes with connective tissue reaction and inhibits the formation of fibroblasts and collagen

The remarkable effect of the drug manifested itself in a striking improvement of the clinical symptoms There was diminution of stiffness, decrease of tenderness and pain on motion, decrease of swelling, return of function occurring, sometimes within a few hours or a few days There was also a decrease of the sedimentation rate

There is hardly any doubt that this is the most sensational remedy against rheumatoid arthritis and allied conditions yet discovered The subsequent responses to the discovery which appeared in great rapidity told essentially the same story striking and rapid improvement

Slocumb, Hench, Polley and Kendall then reported 23 patients with very marked or marked relief in 22

It had already been observed by Hench and by his associates that the effect

of the drug was only temporary, that is, it disappeared as soon as the drug was withdrawn or discontinued They felt that it was necessary above all things to establish a certain maintenance dose in order to secure a more lasting result

In view of the enormous benefit which undoubtedly this powerful remedy afforded to the patient suffering from rheumatoid arthritis and allied conditions, and in view of the enthusiastic reception which the drug has received by the entire medical profession, it is only fair to emphasize that these hormones are capable of producing a considerable amount of undesirable effects which can only be avoided by the greatest vigilance and selection of cases

Hench and his associates had already noticed such effects as hirsutism, the rounding of the face the cessation of menses, keratosis and so forth More serious effects are negative nitrogen balance, an increased excretion of creatin and uric acid, diminution of the blood potassium with muscular weakness, water retention and psychic changes Of particular importance, however, in the administration of Cortisone is the danger of adrenal exhaustion Consequently it is obvious that the administration of this drug must be strictly supervised The drug must be discontinued immediately upon the appearance of any of these threatening signs if one wants to avoid serious consequences [3]

So far as the dosage is concerned, Hench and his associates recommended a parenteric or intramuscular administration of 300 mgm the first day followed by 100 mgm daily thereafter In our own practice we apply 300 mgm intramuscularly the first day, followed by 200 mgm on the second day, and 100 on the third day for a number of days Oral administration is also advised but the dosage required is about 30% higher than the parenteric dosage From our rather small series we can confirm, as a whole, that the striking improvement, which is found either subjectively or objectively or both, occurs often within a few hours or days

However, we also find that there is a rapid return of symptoms when the drug is discontinued and in several cases the remedy had to be abandoned because of the disturbance of the kidney function, or its effect upon the central nervous system Neither have we been able to this date to determine the maintenance dose adequate to insure a permanent improvement

No doubt further study will be needed before the drug is firmly established among the medicinal measures for the control of rheumatoid arthritis

8 Summarizing the value of different medicinal drugs

Summarizing the value of different medicinal drugs in rheumatoid arthritis, the conclusions are as follows *Sulfur* therapy should be rejected, it has no rationale and no effect The vaccines are losing their hold, their effect is largely psychological *Fever therapy* in the form of foreign proteins, typhoid, etc, shows only transitory results there are only a few cases of permanent improvement *Sulfonamides*, as well as *penicillin*, have no value whatsoever *Gold salts* are most promising but they are dangerous because of the frequency of

toxic reaction We favor the use of soluble crystalline salts, that is, gold sodium thiomalate and sodium thiosulfate The best means of administration is intramuscularly, starting with 5 to 10 mg up to 50 mg per weekly dose, to reach a total of 1000 mg in the course of treatment

At the time of this writing *Cortisone* and *ACTH* still dominate the field of antirheumatic remedies in spite of accumulating reports on undesirable and even severe side reactions It seems the difficulty lies now more in finding an effective way of prolonging the effect of the drug by establishing an adequate maintenance level

IV THE GENERAL TREATMENT OF RHEUMATOID ARTHRITIS BY PHYSICAL REMEDIES

A THE APPLICATION OF HEAT

Some forms of heat produce hyperemia of the superficial structures and thereby relieve the deep structures of congestion, particularly the synovial membrane, others have a more penetrating effect

1 Thermotherapy

Whether it is applied in the form of radiant heat by heat cabinets, infra red light, or in the form of contact heat by hydrotherapy, hot baths, wet packs or dressing, or by immersion in the so called Hubbard tank, the effect is similar It causes deflection of the hyperemia to the periphery Only in the convective heat by diathermy and short waves can a real temperature increase of the deep tissues be produced, causing a deep hyperemia which opens the way for the recession of reversible joint changes Unfortunately the acutely inflamed joint tolerates convective deep heat badly, so that often one must be content with contact or radiant heat, which is entirely symptomatic in its effect

2 Fever therapy

It is based upon the idea that the body temperature can be raised to a point where it is bactericidal for the invading organism within a few hours For instance, in gonorrhea such a temperature would be 106 to 107 degrees F In rheumatoid arthritis, however, this raising of temperature does not seem to strike the causative organism at all, even though some report that from 30 to 60 per cent of the patients were improved As this treatment is usually combined with other measures, it is difficult to evaluate Comroe[8] stated (1941) that while the results of fever therapy are excellent in gonorrheal arthritis, they are poor in rheumatoid arthritis The same may be said of fever introduced by malaria We have already discussed the temperature reaction induced by the introduction of foreign protein Neither of these seems to "hit the spot" in the treatment of rheumatoid arthritis

of the drug was only temporary, that is, it disappeared as soon as the drug was withdrawn or discontinued They felt that it was necessary above all things to establish a certain maintenance dose in order to secure a more lasting result

In view of the enormous benefit which undoubtedly this powerful remedy afforded to the patient suffering from rheumatoid arthritis and allied conditions, and in view of the enthusiastic reception which the drug has received by the entire medical profession, it is only fair to emphasize that these hormones are capable of producing a considerable amount of undesirable effects which can only be avoided by the greatest vigilance and selection of cases

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methods of applying heat to produce hyperemia More often than not, the effect is temporary, but it should be mentioned that in other types of arthritis, for instance in the ankylosing arthritis of the spine and in certain cases of hypertrophic arthritis, x ray treatment shows some remarkable results

REFERENCES

1 ABRAMS N R and BAUER, W *J A M A* *111* 1632, 1938
2 BAGLEY W R *Minnesota Med* *28* 205 1945
3 BOLAND E W HEADLEY, N E and HENCH P S *Proc Staff Meet, Mayo Clinic,* October 18 1944
4 BORAK, J and TAYLOR, H *Arch Physiotherap, 45* *377* 1945
5 BROWNING J S, RICE R M LEE W V and BAKER, L M *New England J Med* *237* 428 1947
6 CLARKE W H *Research Seminar Notes,* Dept Orthop Surg, State Univ of Iowa, *18 D* 3 1946 1947
7 COHEN A and GOLDMAN J *New England J Med* *223* Aug 16 1945
8 COMROE B I *J A M A* *128* 848 1945
9 CROWE H W *J Lab & Clin Med* *15* 1072 1930
10 —— *Lancet* *1* 563 1944
11 FARILL J Personal Communication Mexico
12 FORESTIER J *J Lab & Clin Med* *11* 1074 1938
13 GARDNER E R *Med Rec, 153* 321 1941
14 HARTFALL S J and GARLAND H G *Lancet* *1* 1459, 1936, *2* 1410 1938
15 HENCH P S KENDALL E C SLOCUMB C H and POLLEY H F The effect of a hormone of the adrenal cortex (17 hydro 11 dehydrocorticosterone Compound E) and of a pituitary adrenocorticotropic hormone on rheumatoid arthritis, preliminary Report *Proc Staff Meet, Mayo Clin* *24* 181 (April 13) 1949
16 KAHLMETER G *Proc Roy Soc Med* *25* 1117 1932
17 —— *Brit Nat'l Com Chron Rheum Dis Annual Report* *3* 17 1937
18 KLING D H *Ann Surg, 99* 568 1934
19 KROYER J and Others *Ann Int Med* *11* 1077 1938
20 MAGNUSSEN P B MCELVINNY R C and LOGAN C B *J Michigan State Med Soc* *46* 1 1947
21 PRICE A E and LEICHTENTRITT B *Ann Int Med* *19* 70 1943
22 RINEHART J F *et al* *Arch Int Med* *61* 537 1938
23 SIDEL N and ABRAMS M I *J A M A* *114* 1740 1940
24 SLOCUMB C A *Ann Int Med* *16* 241 1942
25 SNYDER R G and SQUIRES W H *New York State J Med* *40* 708 1940
26 WEINBERG T B *Am J Roentgenol* *43* 416 1940
27 WHEELDON T *Ann Int Med* *7* 1540 1934

3 Heliotherapy

One can speak only of the general effect of this modality Sunlight relaxes the muscle spasm and, combined with a good appropriate diet, is an excellent tonic for the general condition, but it has no specific effect on arthritics

B THE SPAS

Mineral springs and other watering places have always held an important place in the minds of the public The best one can say for them in regard to arthritis is that they afford the patient the required rest, relieve him from probably troublesome surroundings, and compel him to lead a comfortable and regular life in which proper diet, outdoor exercise and fresh air can be administered without interference Because in most of these resorts, facilities are at hand for proper physiotherapy, the systemic treatment is supplemented by local application which, of course, is of benefit for the local control of the disease So far as the merits of the springs themselves are concerned, the water is often laxative and aids in the general elimination, even if it does not have any specific virtue in the control of arthritis The greatest value of these springs lies in the purgative action of the mineral waters, as well as the application of heat in the form of baths, packs, mud baths or fango The latter is a mud containing water, organic, and inorganic matter such as calcium, sulfur, silica, iron and magnesium Its effect is a dilatation of the blood vessels which again causes more rapid absorption of the joint effusions and relief of pain

V THE X RAY TREATMENT IN ATROPHIC ARTHRITIS

The principal exponent of the application of x ray to arthritis was Kahlmeter[16] (1932), who reported 180 cases of rheumatoid arthritis, of which 60 per cent were markedly improved The good results consisted in disappearance of pain, better mobility, and disappearance of capsular thickening In 1937 he was able to report over 4,000 cases of all kinds, which he treated by x ray over a period of 10 years[17] A few years later T B Weinberg[6] reported 161 cases comprising bursitis, arthritis and spondylitis, of which 82 were cured and 75 were improved Further reports were made by Borak and Taylor[4] These authors claimed beneficial results in a smaller series of cases of rheumatoid arthritis in which gold and physiotherapy had failed They used dosages of 800 to 1600 R administered in three weeks' periods The beneficial effects were decrease of pain and increase of motion

Results of our experience in 45 cases treated between 1942 and 1945[6] showed that 70 per cent of the cases of atrophic arthritis received some relief from pain but in only 15 per cent of the total did the relief persist

It is difficult to account for the effect of x ray on arthritics, except on the ground that it produces a local hyperemia which facilitates absorption and other changes reversible in nature If acute symptoms are present, such as swelling and restricted motion due to painfulness of the periarticular structures then the x ray treatment will have some effect, the same as other

A THE PRINCIPLE OF REST

In the acute stages the function of the joint must be suspended If it is a weight bearing joint, it must be relieved of this function, the joint must be placed in midposition, in which the synovial lining is under least tension

Positions of election For the knee joint it is semiflexion, for the ankle joint, a right angle position is normal, avoiding equinus contracture, for the hip joint, a position of slight flexion and neutral rotation, for the shoulder joint, a position of slight abduction, for the elbow joint, a position of 70 to 80 degrees flexion, midway between pronation and supination, for the wrist joint, a position of 10 degrees dorsal extension with slight flexion in the meta carpophalangeal and interphalangeal joints Immobilization of the joints in this position is usually carried out by splints or bandages, hardly ever by plaster

How long is the immobilization to be continued? In arthritics it should not extend beyond the painful period It should be abandoned definitely when swelling and pain have subsided enough so that the patient himself can carry out active movement During the acute stages, splints should be removed temporarily to permit physiotherapeutic measures to be applied

B LOCAL PHYSIOTHERAPY FOR CONTROL OF PAIN AND SWELLING

1 Local application of heat

Heat is used in various forms As radiant heat from an infra red lamp, as contact heat in form of hot towels or packs, as penetrating heat by diathermy or short wave radiation The choice of the modality depends, naturally, upon the status of the joint Very sensitive joints will not tolerate penetrating heat such as is furnished by diathermy or short wave radiation When the joint becomes tolerant to any kind of heat, that form which provides the most superficial hyperemia is best to control pain The application should be made once or twice daily, the splints can be removed for the purpose, and the joint placed in a comfortable position

2 Local hyperemia for control of pain and swelling

In the description of the general treatment, it was mentioned that vasodila tory drugs like histamine and mecholyl are used to produce a general dilata tion of the blood vessels Attempts have also been made to provide for increased peripheral circulation in a more local sense by operative means Adson and Rowntree[1] recommended sympathetic ganglionectomy to bring about increased circulation in the affected joints The first report of Adson included five favorable cases It was later substantiated by J A Moore[9] The effect of the operation is, of course, entirely circulatory, but there is an im provement in the function of the joint inasmuch as pain and swelling are reduced A later report (A Young[17]) describes six cases treated by sympa

Lecture III

ON THE ORTHOPEDIC TREATMENT OF ATROPHIC ARTHRITIS

OVER one's efforts to restore the patient's resistance and to elevate his natural immunity, one should not lose sight of the requirements of the local situation That this happens only too often is manifested bv the multitude of deformities and contractures permitted to develop beyond the point where preventive or corrective measures would be of avail Transitory or reversible changes can be handled by such restorative methods as will facilitate absorption of infiltrates or exudates in the early stage of the disease Permanent changes, on the other hand, require reconstructive methods which are at best only substitutes for natural function

The implication is that the surgeon must have a clear conception of the pathological background so as to be able to distinguish symptoms which are based on reversible changes from those which are due to irreversible ones The best results naturally are obtained by conservative methods They do not interfere with the anatomy and physiology of the diseased joints, however, their usefulness ends when permanent pathological changes appear

In general terms, the objectives of the local treatment can be formulated as follows

1 The control of swelling and pain
2 The prevention and treatment of muscular atrophy and joint instability
3 Prevention and treatment of contractures
4 The restoration of weight bearing tolerance
5 The restoration of stability of the affected joints
6 The correction of fixed deformities in ankylosed joints
7 The restoration of mobility of the joints

One must expect a limit to conservative treatment in respect to each of these objectives Nevertheless, we know from experience that the possibilities for conservative treatment are much greater than they are generally given credit for At any rate, the time for it is the period of reversible joint changes As the disease becomes more chronic and the changes more definite, conservative treatment gradually recedes into the background

I THE CONTROL OF PAIN AND SWELLING BY LOCAL TREATMENT

Sedatives and anodynes which are given to the patient in the earliest stages are to be considered as stopgaps only Control of pain should be achieved by local means

trol Weight bearing tolerance plays a most important role in the entire scheme of treatment Obviously, during the most acute stage the patient is bedfast, but as soon as that stage has passed recumbency should not be prolonged The upright position must be assumed as soon as possible, and the patient should be helped and encouraged to leave the bed

Should braces be applied for the support of weight bearing articulations? As a temporary measure, they are fully justified, but they should remain only until the muscles have become strong enough to withstand static and weight-bearing stresses When the patient leaves the bed the program for the lower extremities includes the wearing of supportive braces with free knee joint and ankle joint, to make active motion possible in these joints At the same time the arches of the foot must be protected, because the foot muscles are so lacking in tone that temporary support by insoles and shoe corrections becomes necessary During all this time, massage and exercises and other modalities of physical therapy must be applied systematically The resumption of the function of the upper extremity is initiated at the earliest possible moment by massage and active motion This is followed by occupational therapy, to develop directed and independent motion in all articulations, particularly the wrist and fingers

III THE PREVENTION AND TREATMENT OF CONTRACTURES IN ATROPHIC ARTHRITIS

A GENERAL CONSIDERATIONS

The contractures of the earlier stages of arthritis are the result of reflex spasm Because of their spastic nature, they are eminently amenable to mechanical correction During this early period, positional contractures develop in the lower extremity, particularly in the ankle joint, due to the relative weakness of the extensors The prevention of the contracture is a matter of positioning when the patient is in bed and proper splinting when he is ambulatory

In the later stages of the disease, these contracture positions lead to structural shortening of the muscles with definite histological changes The result is that muscles become fixed and no longer yield to passive stretching At this stage operative methods become necessary for correction These operative procedures have the disadvantage that they interfere with the normal function of the muscles by weakening their contractile strength It is therefore of prime importance to prevent contracture, or at least to recognize its development in the earliest stages

In the upper extremity two types of contractures are prevalent, namely, the clawhand and the pill roller hand A tendency to either type can be recognized very early by a certain rigidity of the muscles and by the resistance of the joints to passive motion The elbow joint contractures occur in position of flexion and pronation the shoulder joint contractures, in adduction In the lower extremities, we must expect flexion contractures of the hip and knee,

thectomy of the 2nd to 4th lumbar ganglion, with resulting improvement in motion and loss of pain There is a good rationale behind this treatment It is a fact that the increased hyperemia which extends through the entire length of the limb and involves the joints as well as the other soft tissues is favorable for the absorption and gradual disappearance of reversible exudates or infiltrations The patient experiences a definite feeling of well being From the reports mentioned above, the immediate results of this sympathectomy were certainly encouraging If the method has not met with more general acceptance, either for the control of pain or of swelling, it is because it is transitory in its effect After the vasodilatation has receded, which happens sometimes within a few days, the pain and swelling recur, but the immediate effect of the method is proof that local treatment, especially the application of heat with its resulting hyperemia, is quite capable of suppressing pain and controlling swelling in the earlier stages of the disease

II THE PREVENTION AND TREATMENT OF MUSCULAR ATROPHY IN ARTHRITIS

Muscular atrophy proceeds rapidly in the acute stage, with the result that all the weight bearing joints become unstable But this instability is only partly due to the shrinkage of the muscles It is also produced by the strain to which the reinforcing ligaments are subjected from the persistent effusion and distention of the joint

A MASSAGE

The best protection against instability is the muscle tone For this reason, early massage of the arthritic muscles is indispensable How soon should massage be instituted? If we combine it with hot packs or radiation, it can be carried out very early, at least in the form of gentle friction It is also helpful at this stage to use faradization of adequate intensity to keep the muscles in contraction the electrodes are applied to Erb's points

B ACTIVE EXERCISES

Exercises should be started early, it is best to proceed on a definite plan giving special consideration to those muscles which are most essential for stabilization of the joint and for maintenance of the upright position For the ankle joint, this would be the triceps surae, for the knee joint, the quadriceps, and for the hip joint, the gluteals

C RESUMPTION OF STATIC FUNCTION

In the interest of proper muscle development and avoidance of muscle atrophy, it is essential that static function should be resumed as early as possible There are several reasons why arthritics should get out of bed as soon as possible, but the most important is to combat rapidly progressing muscular atrophy and relaxation In order to do that, however, pain must be under con

trol Weight bearing tolerance plays a most important role in the entire scheme of treatment Obviously, during the most acute stage the patient is bedfast, but as soon as that stage has passed recumbency should not be prolonged The upright position must be assumed as soon as possible, and the patient should be helped and encouraged to leave the bed

Should braces be applied for the support of weight bearing articulations? As a temporary measure, they are fully justified, but they should remain only until the muscles have become strong enough to withstand static and weight bearing stresses When the patient leaves the bed the program for the lower extremities includes the wearing of supportive braces with free knee joint and ankle joint, to make active motion possible in these joints At the same time the arches of the foot must be protected, because the foot muscles are so lacking in tone that temporary support by insoles and shoe corrections becomes necessary During all this time, massage and exercises and other modalities of physical therapy must be applied systematically The resumption of the function of the upper extremity is initiated at the earliest possible moment by massage and active motion This is followed by occupational therapy, to develop directed and independent motion in all articulations, particularly the wrist and fingers

III THE PREVENTION AND TREATMENT OF CONTRACTURES IN ATROPHIC ARTHRITIS

A GENERAL CONSIDERATIONS

The contractures of the earlier stages of arthritis are the result of reflex spasm Because of their spastic nature, they are eminently amenable to mechanical correction During this early period, positional contractures develop in the lower extremity, particularly in the ankle joint, due to the relative weakness of the extensors The prevention of the contracture is a matter of positioning when the patient is in bed and proper splinting when he is ambulatory

In the later stages of the disease, these contracture positions lead to structural shortening of the muscles with definite histological changes The result is that muscles become fixed and no longer yield to passive stretching At this stage operative methods become necessary for correction These operative procedures have the disadvantage that they interfere with the normal function of the muscles by weakening their contractile strength It is therefore of prime importance to prevent contracture, or at least to recognize its development in the earliest stages

In the upper extremity two types of contractures are prevalent, namely, the clawhand and the pill roller hand A tendency to either type can be recognized very early by a certain rigidity of the muscles and by the resistance of the joints to passive motion The elbow joint contractures occur in position of flexion and pronation the shoulder joint contractures, in adduction In the lower extremities, we must expect flexion contractures of the hip and knee,-

and equinus contracture of the ankle joint Added to this is the valgus deformity of the foot as the patient becomes ambulatory

Because the conservative treatment of contractures by mechanical stretching is far superior to operative methods, it is most essential to realize to what extent conservative measures can be employed As a general rule one may assume that the limit of conservative treatment is reached as soon as signs of disalignments of the joint appear in form of subluxations In contractures of long standing, not only are muscles shortened, but the capsule and ligamentous structures likewise take part in the shrinkage The latter offer insuperable obstacles to mechanical correction It is at this point that operative methods such as tenotomy, capsulotomy, tendon lengthening, capsular stripping, osteotomy or joint resection must be resorted to

B SPECIAL ORTHOPEDIC MEASURES FOR CORRECTION OF CONTRACTURES OF THE LOWER EXTREMITIES

1 The ankle and foot

a Equinus and Equinovalgus Contractures

The most common deformities are the equinus and equinovalgus contractures The equinus often develops during bed rest, the valgus deformity, mostly when the patient becomes ambulatory The equinus is frequently accompanied by retraction of the toes, a clawfoot develops because the extensor tendons are under tension and the weakened flexor tendons of the toes are no longer able to hold their balance against them

Case J B (Male) #M 13621
Age 63 years Adm September 21, 1936

A case of atrophic arthritis of the left knee and ankle with contractures These started after the patient had been bedfast for six months When he became ambulatory the knee was in good position, but the left foot was in equinus contracture and could not be corrected by manipulation

In the early stages this deformity can be corrected by splints, and later by manipulation, under anesthesia if necessary It should then be followed by application of a plaster cast to hold the foot in neutral position and at a right angle In other cases the correction can be accomplished gradually by a turnbuckle arrangement or by elastic traction (Fig 23) Whether it is obtained by manipulation, elastic traction, or a turnbuckle arrangement or cast, the position of correction must be maintained for at least three or four weeks Physiotherapy



Fig 23 Turnbuckle arrangement for correction of equinus

must then be instituted to develop, by passive manipulation, heat, and exercises, the active and passive mobility of the joints and to enhance the development of the musculature In some instances it becomes necessary to protect the joints by braces for some time after the patient has resumed standing and walking

On the other hand, when a muscle is already structurally shortened and there is retraction and shrinkage of the capsular apparatus, conservative methods no longer suffice Here operative procedures are necessary It is by no means always sufficient to lengthen the shortened heel cord More often than not, a shrinkage of the capsule which occurs concurrently with the retraction of the tendo achillis will require an extensive resection of the posterior capsular structures

b CLAWFOOT DEFORMITY

In earlier stages, this deformity also responds to manipulation, followed by a corrective cast in which the toes are held firmly downward in volar flexion This cast also remains for a period of three or four weeks, after which mobility is maintained by physiotherapeutic measures In the most severe cases where the basal phalanges are actually overriding the necks of the metatarsals, one may have to resect the metatarsal heads according to the procedure introduced by Hoffman Furthermore, in the later stages of the arthritic clawfoot a fixed hammer toe deformity develops Associated with the so called "clawing" in the metatarsophalangeal joint, this fixed hammer-toe deformity requires operative measures which consist of resection of the proximal interphalangeal joint

2 The knee joint

a CONSERVATIVE TREATMENT

The arthritic knee joint contracts in position of flexion and outward rotation In the beginning, this contracture can be prevented by positioning or by splinting It is essential, however, that this position be maintained only temporarily, that is, until the patient is over the most painful stage of the disease At the earliest moment possible, even when the patient is still in bed, a systematic regime of developing muscle tone should be instituted by massage of the quadriceps and other muscles of the legs, by muscle setting exercises, and other types of active motion

Traction devices If the knee joint persists in assuming a contracture position due to spasticity of the hamstring muscles, the best measure to overcome this is to apply traction during the recumbent period The traction must work in the direction of the leg and not of the thigh, if the traction operates in the direction of the thigh and not the leg, the result is a rotatory effect which ends in posterior subluxation of the leg upon the thigh, especially if the ligamentous apparatus is considerably relaxed (Figs 24 and 25)

As soon as the condition of the joint permits active exercises, they should







Fig 24 Russell traction arranged for the knee



Fig 25 Result of traction in straightening arthritic contracture of the knee Case D A

be carried out, while the patient is still in bed A simple mechanical device carrying weights can be used for resistive active exercises

Turnbuckle casts This is a very efficient device for the correction of the more resistant flexion contracture of the knee joint The application requires painstaking technique and a great deal of patience and perseverence, but it is surprising how much can be accomplished by judicious application of a turnbuckle cast on a patient who has the necessary determination (Figs 26 and 27)

The success of the method depends very much upon the manner in which the cast is applied It is an angulated long leg cast which is hinged at the knee joint, the hinges must be placed exactly in the proper anatomical position The cast is then cut out on the underside, leaving only a small anterior bridge

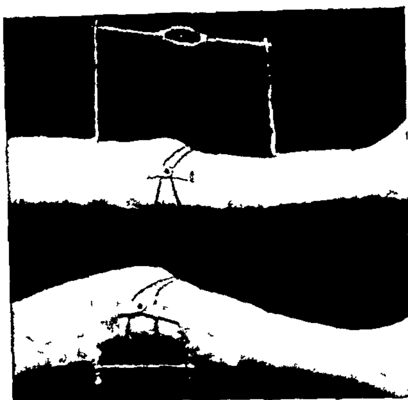


Fig 26 The turnbuckle cast for straightening the contracted knee

Extension is then carried out by means of turnbuckles with hinges applied at both sides of the cast, or by means of elastic bands or rubber tubes applied in front over two angulated irons We find that it is not always necessary nor is it feasible to carry out the correction until full extension is obtained Most of the time it is sufficient to approach the straight position, and the remainder of the deformity can be overcome by simple operative means It is not unusual to find that half a year or more is required before this point is reached After the turnbuckle cast is removed, the knee still must be protected by a brace against recurrence of the contracture J Kulowski,[8] in a review of 44 cases of flexion contracture of the knee treated by the turnbuckle method in our clinic, found the average duration of contracture to be 3 47 years, and the average amount of contracture, 40 degrees Complete correction by turnbuckle treatment was obtained in 36 cases, in an average treatment time of 20 days This short period of correction applies, of course, only to the acute and subacute cases

b Operative Methods for Correction of Contractured Knees in Atrophic Arthritis

The only criterion to indicate that the resistance of the contracted tissue is greater than the stability of the joint itself is the appearance of a subluxation of the tibia on the femur under continued mechanical correction From that point on, operative measures must be carried out The proper sequence is to attack the structures farthest from the joint and then to proceed to those in

the immediate vicinity, that is, the tendons and muscles should be lengthened or tenotomized first, then one proceeds to release the deeper structures, such as the capsular apparatus If, after that complete correction is stopped by the resistance of the nerves and vessels, direct operative correction is at an end, for the remainder of the deformity one must resort to the pseudo correction by supracondylar osteotomy



FIG ?, Extreme arthritic flexion contracture corrected by turnbuckle cast and hamstring tenotomies Case E G (female) #61 939 49 years August 1947 Patient had been bedfast for 11 years Knees in acute flexion and heels touching buttocks Correction by 10 months of turn buckle casts followed by hamstring tenotomies Was discharged walking a) Before b) After treatment

The hamstring tenotomy The tendons can be lengthened or tenotomized by either a median incision which runs through the middle of the popliteal space or by two separate incisions, one medial and one lateral In the latter case the incision for the biceps is made directly over this tendon, and that for the inner hamstring is made over the projecting tendon of the semi tendinosus

The posterior capsulotomy Here the operative plan is the stripping of the proximal insertion of the joint capsule from the posterior surface of the femur We use the technique of P D Wilson [16] The capsule is reached by two incisions, one on the outer side, extending from the middle of the lateral aspect of the femur just above the condyles to the head of the fibula, and one of the same length on the inner side, from above the adductor tubercle on the medial side downward slightly below the joint line At the outer side, the iliotibial band is sectioned and the biceps is split, then the lateral portion of the capsule of the knee joint is uncovered and stripped down from the back of the femur A similar procedure is used along the inner side

Osteotomy After all the soft tissue obstacles have been overcome by tenotomy of the hamstrings and by the stripping of the capsule, one may find that the popliteal vessels and nerves become tight on further attempt to straighten the knee This sets a halt to the direct correction of the deformity It is then possible to get further correction by a supracondylar osteotomy which produces a counter kink above the knee joint, offsetting the remaining degree of knee joint contracture Even then, one must be careful that the correction does not place undue tension upon the neurovascular structures

Case E M (Female) #41 11166
Age 44 years Adm August 30, 1941

History of arthritis for eight years and inability to walk for two years On admission the arthritis was quiescent, but the left knee was contracted at 110 degrees, the right knee was fixed at 90 degrees

The treatment of the knees was by turnbuckle casts which straightened the right knee from 90 to 135 degrees, and the left from 110 to 140 degrees To obtain complete correction a supracondylar osteotomy was performed on the left knee five months later The same procedure was then repeated on the right knee in May, 1942 When seen last, in July, 1944, she was able to walk without crutches

3 Correction of hip joint contractures in atrophic arthritis

a CONSERVATIVE

The contractures of the hip joint are much more resistant to mechanical correction than those of the knee, because the uniarticular flexors of this joint undergo structural shortening much sooner If the contracture is to be corrected by traction, it is necessary to immobilize the lumbar spine by a plaster spica, including the pelvis and the opposite thigh

b OPERATIVE

Up to a certain degree, the contracture yields to mechanical stretching by traction or by corrective casts On the whole, however, one will find that more severe hip contractures fail to yield to conservative means Then the question arises whether releasing the contracted muscles from their point of insertion is adequate for correction The operation of Soutter[13] and its later

modification by Campbell[3] are based on this principle The muscles which must be released are the sartorius, the tensor fasciae latae, the rectus femoris, the illiopsoas, and the anterior portions of the gluteus medius and minimus Still, except in moderate contractures, the operative result is only partial, the more severe flexion contracture requires the osteotomy of the femur at the subtrochanteric level Following the osteotomy the hip must be immobilized in a hip spica in corrected position for at least two or three months

One may ask why the osteotomy should not be done first, without previous procedures on the soft parts We believe that it is a good plan to accept the osteotomy as the principal and, in many instances, the only operation It certainly simplifies the problem The stripping is a rather extensive operation and is likely to lead to recurrences because of massive scar formation Besides, it is not free from danger, since the attempt to extend the hip joint forces the pelvis into sharp inclination so long as the joint shows any resistance to extension This danger is avoided by the subtrochanteric osteotomy

If the hip is ankylosed the best position would be from 15 to 25 degrees of flexion, varying with the patient's occupation, but one must first be sure that the lumbar spine has enough mobility to compensate for this degree of flexion If the lumbar spine is movable, then the flexion contracture may be corrected fully, but if the mobility of the spine is restricted by arthritic changes the position must be so calculated that it can be maintained in standing as well as in sitting without compensatory movement of the lumbar spine In this case, the position of 15 to 25 degrees flexion is best

C CORRECTION OF CONTRACTURES OF THE UPPER EXTREMITIES IN ATROPHIC ARTHRITIS

1 Contractures of fingers and metacarpals

This is, without doubt the most neglected situation in the entire field of arthritic deformities Too many hopelessly deformed and distorted hands are carried around by the unfortunate patients, a living reproach to the surgeon who has been neglectful in preventing the deformity or in administering proper treatment

We have mentioned the two principal types of deformity of the hands and fingers They are 1 The clawhand in which the metacarpophalangeal joints are hyperextended and the interphalangeal joints are flexed 2 The so called pill roller hand in which the metacarpophalangeal joints are flexed and the interphalangeal joints are extended

The reason why one deformity appears in one case and a different deformity in another is not clear It depends greatly upon the primary deformity of the wrist Should the wrist go into acute flexion contracture, it is more likely that an extension contracture of the metacarpophalangeal joints and a flexion contracture of the interphalangeal joints will develop This happens because the extensors of the fingers are under strong tension The result is a hyperextension

in the metacarpophalangeal joints, which in turn puts the finger flexors under tension and causes the interphalangeal joints to assume a flexion contracture

On the other hand, in cases in which a flexion contracture develops in the metacarpophalangeal joints, the muscle balance is overthrown by the spastic condition of the intrinsic muscles of the hand, the effect of which is flexion in the metacarpophalangeal and extension in the interphalangeal joints

a Conservative Treatment of Arthritic Contractures of the Hand and Fingers

One should be able to recognize at an early stage which type of deformity the arthritic hand will have a tendency to assume In most cases of metacarpophalangeal extension contracture, conservative treatment, if instituted early enough, is sufficient, certainly as long as no actual subluxation of the basal phalanges upon the metacarpals exists, as is the case in the later stages However, shrinkage of the posterior capsular apparatus of the metacarpophalangeal joints occurs rather soon, and if conservative treatment is not instituted promptly and persistently it may cause considerable difficulty Because of the eminently spastic nature of these contractures in the early stages, we can expect them to respond to consistent mechanical treatment We prefer the adjustable splint in which traction is applied by elastic pull (Fig 28)

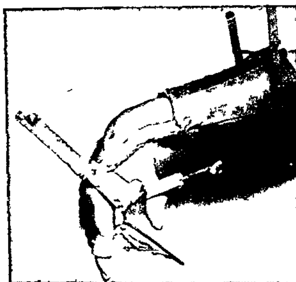


Fig 28 Adjustable splint to overcome flexion contracture of wrist with attachment for extension contracture of midphalangeal joint and adduction contracture of thumb

There are certain mechanical principles to be observed in the use of splints First, each splint should accomplish one thing only A splint which is applied to overcome flexion contracture in the metacarpophalangeal joints must have a basic portion which consists of a cockup splint to which a banjo ring is attached From this banjo ring elastic traction is applied by means of loops which are slipped over the basal phalanges The second mechanical principle is that the correcting force must apply at right angles to the axis of the peripheral portion of the contracted joint, not in line with its axis To provide stability to the corrective force, the metacarpal must be securely immobilized by a bridge resting under the metacarpal necks It is then possible for the elastic traction to act upon the basal phalanges in the proper direction Again, if one attempts to correct a flexion contracture in the interphalangeal joint, it is the basal phalanx which must be stabilized, and the traction must be applied at a right angle to the midphalanx In the flexion contracture of the metacarpophalangeal joints, as seen in the pill roller hand, traction is applied to the basal

phalanges at a right angle and is directed dorsally (Fig 29) In extension contracture of the interphalangeal joint which occurs in combination with flexion contracture of the metacarpophalangeal joint in the pill roller hand type of contracture, the problem becomes more difficult The mechanical principle of correction is the same, namely, the traction is applied at a right angle to the distal portion of the joint, while the proximal portion is held immobilized by a cockup splint provided with a banjo arrangement Here it is somewhat more difficult to secure immobilization of the basal phalanges It is obvious that in this case the force applied at a right angle upon the midphalanges must be directed downward, in opposite direction to that applied for correction of the flexion deformity

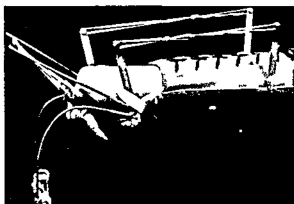


Fig 29 Banjo type splint to overcome flexion contracture in metacarpophalangeal joints

Another rule is that immobilization of finger joints should never be continuous Alternation of splints is important as finger joints become stiff if they are held in one position For this reason we apply splints only for certain hours during the day and make sure that mobilizing procedures are applied during the intervals It must be realized that the wearing of the splint is only an episode in the treatment of the contracture It is absolutely necessary that systematic active and passive motion be carried out constantly during the treatment

If contractures no longer yield to elastic traction, is forcible manipulation permissible? As a general rule, we should warn against so called forcible manipulation because it means intra articular hemorrhage and effusion, and subsequent capsular shrinkage Passive stretching should never be carried to the point of tendinous or capsular tears It must always be gentle, and one must be sure that the elastic "give" of the connective tissue structures is not exhausted

b Operative Methods for Correction of Finger Contractures

The most conservative of these interferences is the capsulotomy of the metacarpophalangeal joint In extension contractures it is the posterior capsular apparatus which is in the state of shrinkage, and in the flexion contracture, the *anterior* *This procedure does not seriously interfere with the function* One will find that correction of the flexion contracture is much more difficult The collateral ligaments of the metacarpophalangeal joints are slack in extension, but they are tight in flexion due to their extensive attachment to the sides of the heads of the metacarpals Dorsally there are no particular reinforcements of the capsular apparatus of the metacarpophalangeal joint, because the dorsal extensions of the extensor muscles effectively serve this purpose But it is an

teriorly where the capsular apparatus is heavily reinforced by fibrocartilaginous plates, the so called volar accessory ligament Consequently, the correction of the deformity by capsulotomy is easier in the extension than in the flexion contracture of the metacarpophalangeal joint

Should one lengthen a contracted extensor or flexor tendon? Such a procedure must be considered carefully Any tendon lengthening invariably weakens the muscle strength because it destroys the natural proportion between tendon length and length of the muscle belly Besides, such a procedure of lengthening of contracted flexor tendons would be unthinkable within the territory of the fingers themselves If any lengthening is to be performed, it must be either in the palm or at the wrist Before it is undertaken one must be sure that there are no other obstacles to the extension of the interphalangeal joint If one finds that the flexion contracture in the interphalangeal joint becomes less as the metacarpophalangeal joint is brought forcibly into a flexion position, it is an indication that the gliding of the tendons in these joints is unimpaired Otherwise we may be sure that other obstacles exist, particularly capsular shrinkage or even intrathecal adhesions Finally, there are extreme cases of clawhand deformity in arthritis where the basal phalanges are strongly over riding the necks of the metacarpals In extreme cases we have resorted to resection of the metacarpal heads in order to save some function of the hand, an operation similar to that which is performed in the extreme clawfoot condition, the so called Hoffman's operation

2 Wrist joint contractures

a Conservative Treatment

The flexion contracture of the wrist is more frequent in arthritis than in any other condition In earlier stages a great deal can be accomplished by proper splinting and daily treatment with heat, passive motion, and massage Between treatments, the hand must be held in position by a cockup splint

b Operative Treatment

If the contracture persists and the muscles have undergone structural shortening, we will find that the flexors of the wrist proper, namely, the flexor carpi radialis and ulnaris, form the principal obstacles They are practically uniarticular muscles and therefore undergo structural shortening more easily than do the pluriarticular finger flexors These flexors may be lengthened because the disadvantage of weakening the muscle force by lengthening the tendon is outweighed by the advantage of giving the wrist the functionally better position of extension However, before such an operation is performed on these muscles, one must be sure that the fingers can be straightened out in the interphalangeal joints, so that when the wrist is returned to normal position the full benefit of the operation can be obtained

3 Elbow joint contractures

a Conservative Treatment

Contractures of the elbow joint are usually in flexion In the early stages splinting and careful passive stretching will give results, all kinds of devices may be used for this purpose, for instance, turnbuckle braces or elastic traction (Fig 30)

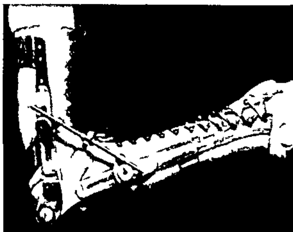


Fig 30 Turnbuckle brace for flexion contracture of elbow

The extension contractures of the elbow are less frequent, but they are much more difficult to handle Prevention of this contracture is so much more imperative, as an elbow ankylosed in extension renders the extremity practically useless

b Operative Treatment

Rarely are operative methods applied for correction of these contractures The only procedure which we have carried out in selected cases was lengthening or tenotomy of the biceps or brachialis anticus muscle

4 Contractures of the shoulder joint

The usual contractures are in inward rotation and adduction Passive stretching and position is sufficient in the earliest stages Difficulties arise when there is capsular shrinkage together with structural shortening of the contracted muscles We warn against brisk manipulation of the shoulder joint in cases of capsular shrinkage, as it almost certainly leads to injury of the musculotendinous cuff and may occasionally even produce fractures of the neck of the humerus

IV THE RESTORATION OF WEIGHT BEARING TOLERANCE IN THE ARTHRITIC JOINT

This is the fourth objective of the local treatment of arthritis Weight bearing tolerance does not develop easily in the large joints of the lower extremity First, it is the activity of the arthritic process itself which makes weight bearing pressure intolerable Then, secondary deformities develop in the course of arthritis which render weight bearing and walking mechanically difficult As a general rule, weight bearing should not be forced upon the patient as long as his joints are not fully tolerant to it It is essential that deformities and contractures be corrected before any kind of weight bearing is undertaken

A THE ANKLE AND TARSAL JOINTS

The prerequisite for weight bearing tolerance is the correction of any deformity The ankle joint and the tarsus must be kept immobilized in corrected

position until both spontaneous pain and swelling have disappeared completely Weight bearing is then instituted gradually, and the patient is kept on crutches until full tolerance is restored Support of the ankle by a leather insole, a leather anklet, braces, or a T strap iron will be necessary for a time

B THE KNEE JOINT

We find that the knee joint rebels against weight bearing more than any other articulation in the lower extremity There are several reasons for this In the first place, the shrinkage of the extensor muscles of the knee is so rapid that the joint loses a great deal of its natural stability In the second place, the changes within the joint itself, particularly the synovial hypertrophy and the enlargement of the fat pads, cause internal impingements The patient avoids full extension of the knee, and walks with his knee slightly flexed

1 Conservative measures to obtain weight bearing tolerance in the arthritic knee

The first attempts to bear weight should be made with the limb protected by some apparatus such as a knee cage, or better still, a full length brace with a movable joint at the knee and ankle and a foot plate

2 Operative procedures for restoration of weight bearing ability of the knee joint

SYNOVECTOMY

If we speak of synovectomy as an operation for the restoration of weight bearing ability, it is because we believe this to be its principal effect The original idea behind this operation was to remove the synovial membrane as a focus of infection, and thereby to control the arthritis itself From this point of view the operation is not justified There is no indication that synovial membrane resection has any effect on the arthritic condition as such On the other hand, by removing the mechanical obstacle of a hypertrophied synovial membrane, a great deal is added to the weight bearing tolerance of the knee Synovectomy does not actually increase the range of motion On the contrary, it often decreases it, but it enables the patient to bear weight without pain The first report on this operation performed for acute infectious arthritis goes back to Albertin (1895) At the turn of the century Goldthwait reported 38 patients on whom he removed masses of fibrin and fringes Cases of synovial membrane capsulotomy were mentioned by J B Murphy[10] (1916), but it was not until 1923 that Swett[15] published the first report of this operation's being performed in chronic atrophic arthritis Ellis Jones[6] mentions only 12 cases which he selected from 300 cases of polyarthritis In his report of 1936, W Hetzar[5] stated that he found some cases markedly improved by partial synovectomy, which consisted essentially in the removal of hypertrophied fat pads

A R Smith,[11] surveying 26 of our synovectomies performed on the knee joint



Fig 31 Synovectomy for chronic monarthritis Case C M (female) #53 910 53 years July 1944 Duration of arthritis of left knee four years Total synovectomy synovia markedly fibrosed Walked without pain two months later had full range of painless motion Figure shows massive hypertrophied synovial sac

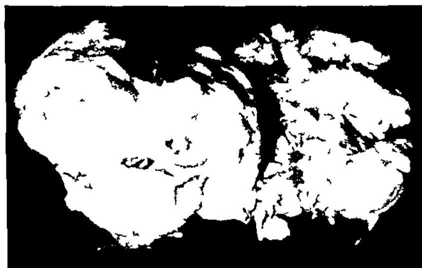


Fig 32 Synovectomy in atrophic arthritis of the knee Case P G (female) #40 10419 44 years August 1940 Pain and swelling of right knee for three years, motion of 20 degrees Continuous pain on weight bearing in spite of physiotherapy and braces Synovectomy in April 1945 following operation had 30 degrees of free motion When seen two years later had minimal discomfort and was able to walk on leg Figure shows hypertrophied synovial sac removed at operation

in chronic arthritis, found that 50 per cent of the monarticular type and only 30 per cent of the polyarticular type gave acceptable results S B Boon Itt, in an analysis of 65 cases of knee synovectomies performed in our clinic on 53 patients, stated that in well selected cases of chronic arthritis of the polyarticular type, 60 per cent were benefited by the method, while of the monarticular type, 75 per cent were improved

If the synovial membrane is partly or entirely removed, how is it replaced? From his clinical observations Swett found that the operation is followed by regeneration of synovial tissue We noticed in experiments on dogs from which we removed the synovial lining of the knee joint, apparently complete restoration in two months, and J Albert Key[7] observed, in reoperated cases, that the joint appeared approximately normal after 60 days

So far as the effect of the operation is concerned, it seems to be mainly mechanical, that is, it improves alignment and weight bearing tolerance In our report of 1925[11] we could show that partial synovectomy, consisting essentially in the removal of fat pads in villous arthritis, has its principal effect on the realignment of the knee These patients do not develop more motion than they had before operation, in our series, some even showed a decided restriction of motion We therefore feel that the removal of soft tissues which prevent complete alignment of the knee joint is fully justified, not because

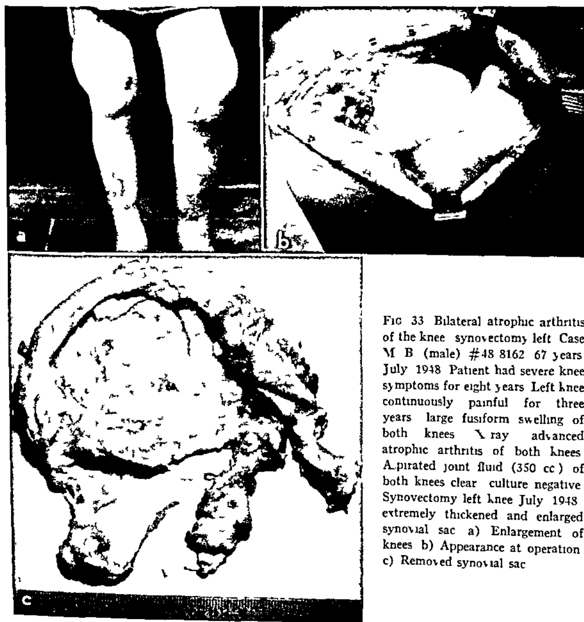


Fig 33 Bilateral atrophic arthritis of the knee synovectomy left Case M B (male) #48 8162 67 years July 1948 Patient had severe knee symptoms for eight years Left knee continuously painful for three years large fusiform swelling of both knees X ray advanced atrophic arthritis of both knees Aspirated joint fluid (350 cc) of both knees clear culture negative Synovectomy left knee July 1948 extremely thickened and enlarged synovial sac a) Enlargement of knees b) Appearance at operation c) Removed synovial sac

it increases motion, but because it produces good alignment and gives a painless weight bearing knee We may, therefore, formulate the specific indications for the operation as follows 1 The condition must be clinically quiescent 2 Synovectomy should be performed in cases where only one or two joints are involved, the knee and possibly another joint Both of these joints should be quiescent (Figs 31, 32 and 33)

C RESTORATION OF WEIGHT BEARING TOLERANCE IN THE ARTHRITIC HIP JOINT

The position of the hip determines, to a great extent, its weight bearing ability If the hip is in marked flexion or adduction contracture, its tolerance is poor for static reasons alone, irrespective of the condition of the joint Osteotomy of the hip is the operation which restores the normal position of the hip

On the other hand, if the intolerance to bear weight is due to the sensitive ness of the joint itself, weight must be eliminated by use of crutches until the joint becomes tolerant There are exceptional cases of atrophic arthritis of the hip in which such tolerance is never established, even though the joint infection seems to have "burned out" Nor does any weight bearing tolerance follow when the position of the joint is corrected In these cases, the solution is fusion of the joint

V THE RESTORATION OF THE STABILITY IN THE ARTHRITIC JOINT

Stability and weight bearing tolerance are not necessarily synonymous Stability is a higher degree of functional integrity To bear weight, one does not need the amount of stability which some joints require for their full function In the hip joint stability in proper position is essential The knee joint, serving as the shortener and lengthener of the limb, requires both mobility and stability in equal measure In the ankle joint, stability in the proper position is the first prerequsite In the upper extremity also, a certain amount of stability is necessary, especially in the shoulder and wrist joints

Even if the shoulder joint is ankylosed, a degree of motion sufficient for practical use may be carried out in the two auxiliary articulations The elbow joint, on the other hand requires free motion because it is the great shortener and lengthener of the extremity In the wrist joint, stability is mechanically essential for the proper action of the flexors and extensors of the fingers The finger joints, on the other hand, require the most motion possible, here stability is a secondary consideration

It may be taken as a general rule that whenever the intrinsic changes in the joint itself have reached a point where natural regeneration is no longer possible, the stability and weight tolerance of such a joint can be obtained only by sacrificing it altogether, that is, either by eliminating it entirely by fusion or by creating a new joint by arthroplasty In some joints the demands made

on stability are much greater than those made on mobility, some joints lend themselves to arthroplastic procedure, while others do not For this reason, the situation must be considered separately for each individual articulation

A THE RESTORATION OF STABILITY IN THE LOWER EXTREMITY

1 The ankle joint

Let us assume that there is no pain in the joint on sitting or in recumbency, but that pain persists on walking and standing and that there is clinical quiescence The decision then rests between the subastragalar arthrodesis and the complete arthrodesis of all joints, including the ankle If the pain appears on walking alone, the subastragalar arthrodesis may be all that is necessary If the x ray picture shows involvement of the ankle joint, and if pain persists during standing as well, it becomes necessary to perform a complete arthrodesis of all articulations The subastragalar arthrodesis should be combined routinely with arthrodesis of the astragaloscaphoid and the calcaneocuboid articulations When all these three articulations, the subastragalar, the midtarsal, and the ankle joint, have to be fused, the proper procedure is the so called panastragalar arthrodesis There are, finally, exceptional cases in which the weight bearing intolerance is confined to the ankle joint alone In these cases fusion of the joint may suffice For this purpose, we prefer the anterior approach which exposes the joint and by forcing the foot into extreme plantar flexion, the greater part of the joint cartilage can be visualized and removed, both from the tibia and from the body of the astragalus

2 The knee joint

Synovectomy procures good alignment and better weight bearing tolerance Its limits are set by the condition of the joint constituents If there is more extensive destruction of the joint cartilage and ulceration of the subchondral bone, the joint does not regain its stability even though all obstacles to align ment have been removed It remains unstable and joint motion must be eliminated altogether In these cases, the fusion is indicated (Fig 34) When both knees are involved, it is advisable to confine the fusion to the one knee which shows the greater degree of destruction

3 The hip joint

Occasionally stability and weight bearing tolerance of this joint cannot be obtained even after an osteotomy is performed for realignment If pain per sists at rest, it is best to perform the arthrodesis (Fig 35) Numerous methods are used for arthrodesing this articulation We prefer a combined intra and extra articular fusion The former consists in a simple ablation of the cartilag- inous surfaces of the acetabulum and of the head of the femur In extra- articular fusion an extra articular bone bridge is constructed from the greater trochanter, as advocated by Hibbs, Hass and many others



FIG 34 Atrophic arthritis of the knee fusion of right knee Case E E (female), #43 6574 6? years September 1935 Arthritis of right knee duration 18 months Swelling and tenderness Fusion *operation* November 1935 marked synovial pannus and extensive erosions Well fused and painless right knee Observation 10 years a) Right knee *before* b) *After* fusion



FIG 35 Atrophic arthritis of the hip *fusion for relief* of pain Case A K (female) #44 1694 56 years August 1944 Hip pain for two years Inability to walk, pain on rest X ray atrophic arthritic changes in hip joint Fusion operation resulting in well fused and painless weight bearing joint ability to walk a) Before b) After fusion

B THE RESTORATION OF STABILITY IN THE UPPER EXTREMITY

Only two articulations are to be considered, the wrist and the shoulder

1 The wrist

Arthrodesis of the wrist is often performed to correct a flexion deformity The operation should leave the wrist in a position of 10 to 20 degrees extension The joint is best approached by a dorsal incision which runs between the tendons of the extensor indicis and the common extensors of the fingers A wedge is removed from the radius and adjoining portion of the scaphoid and semilunar bones or, if necessary, the entire carpus may be removed

2 The shoulder joint

It is only occasionally that an arthrodesis of this articulation is necessary because of persistent pain In performing an arthrodesis, we rely upon the substitutive motion of the acromioclavicular and sternoclavicular articulations However, these articulations have only a limited abduction range For this reason one must be careful not to fuse the arm in too much abduction, otherwise the arm will stand off from the side of the body and will not be capable of complete adduction after the fusion has become solid We have found it advantageous in many cases to add to this procedure the resection of the acromioclavicular articulation, because this gives the shoulder complex a greater abduction range

VI THE RESTORATION OF MOBILITY IN THE ARTHRITIC JOINT

Mobility of stiffened or fused articulations can be obtained only by reconstruction of a new articulation, that is, by arthroplasty, but this operation is applicable to only a few selected joints

A THE LOWER EXTREMITY

There are only two articulations in which mobility can be restored by arthroplasty They are the knee joint and the hip joint

1 The knee joint

When ankylosis of this joint has become established, even if it is only partial and due to fibrous adhesions, any attempt to restore motion by forcible manipulation is not only useless but fraught with considerable danger If the joint destruction has already reached the state of massive adhesions, the joint is beyond natural repair and its motor ability is definitely lost Its restoration can be accomplished only by arthroplasty

An ankylosis due to atrophic arthritis is probably the least favorable condition for arthroplasty of the knee, in comparison with the ankylosis which follows acute septic infection Because of this uncertainty, our indications for

arthroplasty are restricted If one knee only is involved and if it is ankylosed in acceptable position, it should be left alone If both knees are affected, one can hardly avoid operating on one knee, and the choice should be the knee which shows the better bone density and the best endowment of musculature Good musculature is essential in arthroplasty Very often it is advisable to give the patient preparatory muscle training before the operation is performed

The operation requires full exposure of the joint constituents Femur and tibia are then separated with knife or chisel according to the type of ankylosis,



Fig 36 Atrophic arthritis of the knee arthroplasty Case I L (female) #H 27155 19 years February 1933 Pain and swelling in shoulders thumb and left knee with knee remaining ankylosed at 150 degree Conservative treatment to straighten knee of no avail Arthroplasty performed in January 1935 resulting in free motion two years later of 85 degrees from 90 to 175 degrees

whether fibrous or bony The bone ends are shaped with the curved chisel into their natural contours, and a fascial flap is interposed between the tibia and femur (Fig 36)

2 Mobilizing operations of the hip joint

Here the indications are equally strict A hip joint ankylosed in good position should not be a subject for arthroplasty If both hips are ankylosed, however, arthroplasty should be performed on one hip, usually the one which shows more bone density and better musculature (Fig 37)

B MOBILIZING OPERATIONS FOR THE UPPER EXTREMITY

There are only two articulations which are suitable for arthroplasty, the metacarpophalangeal and elbow joints

1 The metacarpophalangeal joints

In extreme contractures where the basal phalanges override the metacarpals, resection of the head with interposition of soft tissue may be performed We have used this operation in several cases, and have restored some mobility However, it must be remembered that this procedure greatly interferes with



FIG 37 Ankylosed hip from atrophic arthritis, arthroplasty Case V W (female) #38 19666 30 years September 1936 Left hip ankylosed in 25 degrees adduction and 30 degrees flexion Anthroplasty of the left hip performed in January 1938 four years later patient had a flexion extension range from 180 to 100 degrees and adduction of 45 degrees a) Before b) After arthroplasty

the stability of these joints and causes considerable weakness of the grip Arthroplasty of the interphalangeal joints in atrophic arthritis did not seem to give us favorable results We used a small vitallium cup for interposition

2 The elbow joint

If this joint is ankylosed, arthroplasty is indicated under certain conditions Where there is extension ankylosis of the elbow, whether complete or incomplete, the operation should be performed, provided the joint is at this time quiescent Should the other joint also be ankylosed, the decision depends upon the position of this joint If it is ankylosed in extension, it also should be subjected to arthroplasty If one joint is ankylosed in flexion and the other in extension, the one in extension should be chosen for arthroplasty

We find, very often that the restriction of motion is only partial and that a certain range has remained The question then is, in what part of the arc does this motion occur The useful amplitude is 40 degrees, the useful arc is between 70 and 110 degrees or between 60 and 100 degrees Smaller ranges

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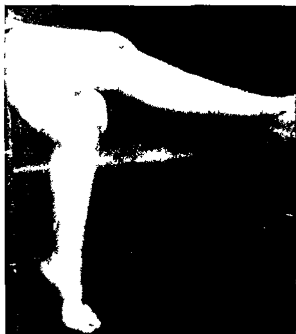


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or more was obtained in only 23 3 per cent All in all, 54 arthroplasties were performed for atrophic arthritis with good results in 38 8 per cent, fair in 31 4 per cent, and poor in 29 6 per cent

REFERENCES

1 ADSON A W and ROWNTREE L G *Surg Gynec & Obst*, *50* 204 1930
2 BOONITT S B *J Bone & Joint Surg*, *12* 853, 1930
3 CAMPBELL, W C *South, M J* *16* 4, 1923
4 HANAVAN E H *Research Seminar Notes,* Dept Orthop Surg State Univ of Iowa *19 D* 13 1948
5 HETZAR W *Arch f Klin Chir* *185* 3, 1936
6 JONES E *J A M A* *81* 1579 1923
7 KEY, J A *J Bone & Joint Surg* *7* 793 1925
8 KULOWSKI J *J Bone & Joint Surg*, *14* 618, 1932
9 MOORE J A *J A M A* *97* 172 1931
10 MURPHY J B *Murphy Clinic* Phila W B Saunders *Vol* V *1916*
11 SMITH A R *Research Seminar Notes* Dept Orthop Surg, State Univ of Iowa *10 D* 37 1936
12 SMITH PETERSON M N *J Bone & Joint Surg* *30 B* 59 1948
13 SOUTTER R *Boston M & S J* *170* 380 1914
14 STEINDLER A *J A M A* *84* 16 1925
15 SWETT P P *J Bone & Joint Surg* *5* 110 1923, *6* 800 1924 Also *Am J Surg* *40* 49 1926 *46* 807 1929
16 WILSON P D *J Bone & Joint Surg* *11* 40 1929
17 YOUNG A *Brit Med J* *2* 375 1936

of motion, especially those which are not in a useful field of motion, leave the elbow handicapped almost as much as a fully ankylosed elbow The principle of the operation is exposure of the joint by means of a posterior incision The joint constituents are dissected subperiosteally and separated, the joint ends are then shaped, so as to resemble more or less the normal contours of the joint, and a fascial flap is interposed The results are, on the whole, superior to those obtained in hip or knee (Fig 38)

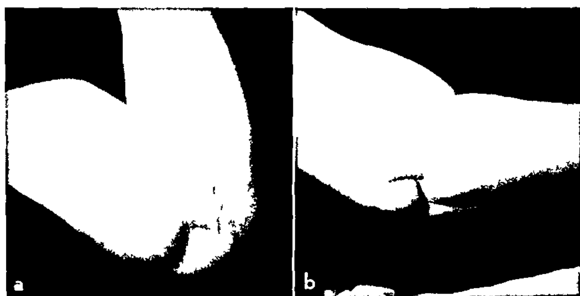


Fig 38 Arthritic ankylosis of elbow arthroplasty Case M K (female) #42 5344 57 years May 1942 Patient had progressive generalized arthritis for 20 years left elbow had flexion range of 85 to 105 degrees and right elbow of 105 to 160 degrees Arthroplasty of the right elbow was performed in November 1944 two years later the range of motion was practically normal

The Statistics on Arthroplasty in Arthritic Joints

The report of Smith Peterson[1] covers 78 cases of arthritis in which arthroplasty of the hip joint was performed, 49 of them bilateral, with a total of 127 operations He states that the results were encouraging, although the range of motion was not as great as in other conditions

Our statistics on arthroplasty of the elbow in atrophic arthritis (Hanavan[4]), including operations performed between 1922 and 1947, are as follows Of 16 arthroplasties performed for atrophic arthritis, excellent results, that is, a flexion extension range of 90 degrees with a good and strong painless joint and good pronation and supination, were obtained in 4 cases, or 25 per cent Good results, with a flexion extension range of 50 to 90 degrees, and 20 degrees or more of pronation and supination, were obtained in 5 cases, or 30 per cent Fair results, with a flexion extension range of 30 to 50 degrees, with active motion and some degree of pronation and supination were obtained in 3 cases, or 18 75 per cent These give a total of excellent, good and fair results in 12 cases, or 75 per cent In the knee joint, good results with 40 degrees or more of motion were obtained in 33 per cent In the hip joint, a range of motion of 40 degrees

C THE STATIC AND OCCUPATIONAL FACTORS

In contrast, static and occupational strains have a decided influence, particularly so far as the weight bearing joints are concerned The resistance to static, occupational, and traumatic factors is lowered, and the joint responds with denegerative changes Evidence of the effect of occupational strain can be seen in the arthritic knee of the seamstress, the arthritic thumb of the cloth cutter, the arthritic shoulder of the carpenter, or the arthritic wrist or elbow of the baseball pitcher Similarly, arthritic changes of the hypertrophic type develop in the elbow and wrist joints of men who use compressed air tools Miners, farmers and factory workers whose joints are heavily taxed in their work constitute a large contingent of arthritics

In addition, there are static disalignments, congenital or acquired, which increase the strain and stress upon the joints and result in degenerative changes when the structures become less tolerant in later years It was Preiser in Germany and Goldthwait in this country who first emphasized the importance of static and postural defects upon the articulations of the body and the connection with hypertrophic arthritis of such deformities as genu valgum and genu varum, or congenital subluxation of the hip joint This is a strong argument for the correction of any static deformity, particularly genu valgum or varum following trauma or due to constitutional diseases such as rickets

D THE TRAUMATIC FACTORS

Inherently the effects of static factors and of acute traumatic events are similar The only difference is in degree and rate of development The so-called traumatic arthritis follows a more intensive accident, which is severe enough to injure the articular surfaces directly Any trauma to a joint resulting from a fracture or dislocation or from a contusion will ultimately produce changes of a degenerative nature, such changes are commonly seen in the subastragalar joint after fracture of the os calcis The regenerative power of the articular cartilage is limited Once a penetrating fracture causes a rent in the joint surface and destroys its smoothness, degenerative changes will result

III THE PATHOLOGY OF DEGENERATIVE ARTHRITIS

A CHANGES IN THE JOINT CARTILAGE

The cartilage is the starting point for all degenerative changes in hypertrophic arthritis It was Pommer[18] who first described the early cartilaginous changes The ground substance of the cartilage begins to appear granular and watery, it then becomes yellowish and discolored, losing its shine and luster, and finally becomes soft and atrophic (Fig 39)

The early microscopic changes consist in fibrillation Fissures and splits are

Lecture IV

ON HYPERTROPHIC ARTHRITIS

I ORIENTATION

SEVERAL terms are attached to Type II of nonspecific arthritis Garrod called it osteoarthritis, a name which still prevails in English literature Virchow's term osteoarthritis deformans is still commonly used in German literature The name hypertrophic arthritis was applied by Goldthwait on the grounds that the enlargement of the joint and the formation of bone were the outstanding features A more descriptive term which stresses the degenerative nature of the disease is hypertrophic degenerative chondro ostoarthritis, intro duced by Weil Nichols and Richardson[16] (1909) used the term degenerative arthritis

II THE PATHOGENESIS

Regarding constitutional tendency the situation parallels that in atrophic arthritis, except that the hypertrophic type is more common in the higher age groups, consequently, the constitutional element of tissue wear and degeneration is more in the foreground Infection is of less significance in the hypertrophic type, but metabolic and static factors as well as traumatism seem to play a more important role

A THE EXOGENOUS INFECTION

Metastatic infection as a cause of hypertrophic arthritis cannot be excluded, even though it is rather unusual Only in 8 per cent of our series of 1300 cases of this type was a focus of infection found, and its removal was followed by sufficient improvement to indicate a direct or indirect connection with the existing arthritis The tonsils seem to play a lesser role than do the teeth and the paranasal sinuses

B THE METABOLIC AND ENDOCRINE FACTORS

On the other hand, it is not unlikely that endocrine factors have some connection with degenerative arthritis For instance, the hypertrophic arthritis of the knee, or the so called Hoffa's disease, occurs most frequently during menopause It is also probable that certain metabolic dysfunctions, particu larly those that have to do with the gastro intestinal tract, have an effect upon the arthritis, but whether this effect is the result of certain toxins from undigested food, or whether it is indirectly due to the lowered general re sistance of the patient is a question Vitamin deficiencies which are important in atrophic arthritis seem to be of lesser consequence in this type

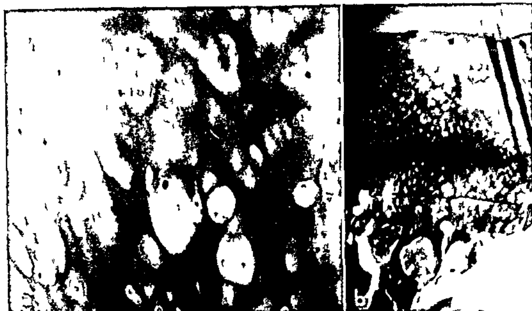


Fig 41 Hypertrophic arthritis Case A S a) Advanced stages of degeneration of joint cartilage granules and watery ground substance also Weichselbaum s lacunae b) Hyper trophic arthritis secondary to a eptic necrosis of the femoral head after a fracture which was secondary and due to mild trauma Penetration of marrow tongues into the zone of calcified cartilage



Fig 42 Hypertrophic arthritis Perforation of sub chondral plate by marrow tongues and extensive ossification of the deep layer of joint cartilage Case F G



Fig 43 Degenerative arthritis Marked degenera tion of joint cartilage perforation of subchondral plate by fibrous marrow In the superficial layers the joint cartilage become fibrocartilage Case F G



FIG 39 Early stages of degenerative arthritis after subchondral fracture of the femoral head Cartilage ground substance appears watery and granular some proliferation of cartilage cells in the pressure zone and some fibrillation

seen in the superficial layers, which finally become detached by friction [13] Eventually this fibrillation reaches the deeper layers, and perpendicular fissures begin to appear The lacunae in which the cartilage cells lie become enlarged, several cartilage cells conglomerate in them and form what is called Weichselbaum's lacunae (Fig 40)

After fibrillation of the cartilage has begun, we see in its deeper layers calcium like deposits in the form of fine granules, which stain dark with hematoxylin The intracellular cartilage matrix shows hyaline and *mucoid degeneration* One notices pits and depressions in the cartilage, which becomes thinner in the weight bearing areas On the other hand, in the marginal portion there appears a different process, namely that of revitalization We see here that the bone marrow of the subcortical layer becomes active again and sends tongues of marrow into the provisory calcification zone where it instigates a reactivation of the enchondral ossification (Figs 41 and 42)

The result is that the center of the joint shows thin and degenerated cartilage which finally disappears completely, whereas the marginal portion not subjected to weight bearing shows a reactivation of enchondral growth In the normal joint the cartilage is entirely quiescent In the arthritic joint, the marginal portion becomes revitalized and undertakes renewed activity As Pommer[18] pointed out, this is mediated by the penetration of marrow vessels and marrow tongues through the calcified zone into the calcium free cartilage (Figs 43 and 44)

B THE MARGINAL EXOSTOSES

This invasion of the marrow tongues beyond the calcification zone leads to the formation of enchondral bone and forms the so called marginal exostoses They are spurs and ledges, found particularly massive in the hip joint where they appear around the head of the femur or the rim of the acetabulum



FIG 40 Degeneration and fibrillation of cartilage Weichselbaum s lacunae

and cyst like cavities develop (Fig 46) Under the effect of pressure, the bone crumbles, and detritus consisting of small particles of necrotic bone fills these cavities (Trummer cysts) At any rate, the cartilage disappears in the center of the joint in the weight bearing areas, and bare bone is exposed Then, under the effect of mechanical friction and pressure, a reactive hypertrophy of the subchondral bone develops, activated by the marrow osteoblasts The denuded bone becomes completely smoothed off under the influence of friction, showing the so-called polishing surfaces Underneath these polishing surfaces one can still see some remnants of cartilage islands in the cancellous zone, as encapsulated cystic formations

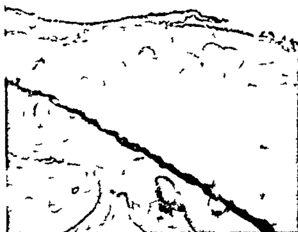


Fig 47 Combined atrophic and hypertrophic arthritis Fibrous tissue pannus covering the joint cartilaginous margin great cartilaginous cell proliferation with formation of Weichselbaum's lacunae Case F G

C THE SYNOVIAL CHANGES

No real inflammatory round cell infiltration is seen at this stage in the synovial membrane The synovial hyperplasia of the villi comes later, and is entirely secondary Consequently there is no pannus formation in hypertrophic arthritis except when it is combined with the atrophic type (Fig 47) The connective tissue structure and reinforcing ligaments of the capsular apparatus show secondary fibrosis and scarring, and it is not uncommon to see metaplastic bone formation included in the capsular apparatus (Fig 48) Because of the fact that the joint cartilage does not become eroded as it does in atrophic arthritis, a



Fig 48 Capsular changes here the joint capsule shows scarring and intracapsular bony tissue formation Case L B



FIG 44 Degenerative arthritis Fibrillation of cartilage cartilaginous cell proliferation and mild calcification of deep layers of cartilage Subchondral bone sclerosis fibrous and adipose marrow Note the fibrous marrow penetrating the subchondral plate in a few areas Case F G



FIG 45 Hypertrophic arthritis and aseptic necrosis of subchondral bone Note fibrosis of bone marrow Case H S

Frequently one finds in the subchondral portions of the marginal exostoses inclusion of cartilage islands, calcified or noncalcified They are cartilage areas which have been by passed by osteoblasts from the bone marrow in their haste to produce new bone

Nichols and Richardson[17] express the opinion that these marginal exostoses come from a compensatory hyperplasia of the bone forming cells of the periosteum This is not correct, it has been proved definitely by Pommer[18] that these formations start from the subchondral bone marrow as it penetrates into the cartilage, and the cancellous core of these exostoses is still covered throughout by the remaining layers of superficial joint cartilage The bone marrow reacts with fibrosis



FIG 46 Subchondral cyst

In traumatic cases a primary aseptic necrosis of the bone itself may be the cause of the fibrosis (Fig 45) As a rule, however, the fibrosis of the bone marrow occurs in response to the changes in the cartilage and is associated with increased activity of the osteoclasts, so that absorption takes place in the subchondral bone at an accentuated rate

nodes (Fig 49) These nodes are tender swellings around the articulation and are due to the thickening of the periarticular tissue They do not represent actual exostoses

B THE LOCAL SIGNS

1 Early symptoms

Early signs are local articular pain, stiffness of the joints, paresthesias, tingling and numbness, followed by limitation of motion At first, the stiffness of the fingers lessens upon movement and exercise Later, pain occurs on motion and in the lower extremities on weight bearing Occasionally subcutaneous nodes are seen at end phalangeal and midphalangeal joints

2 Later and secondary symptoms

a CONTRACTURES

Secondary symptoms are the contractures of the joints due to muscular spasm and result in restriction of motion

b STRESS AND STRAIN SIGNS

The periarticular structures take part in the degenerative process and lose their natural elasticity In the weight bearing joints, there soon appear signs of stress and strain due to involvement of extra articular structures Sometimes actual tears of the periarticular structures occur, as noted particularly in the shoulder and knee joints

c PAIN

The most consistent sign is pain It should be carefully analyzed, as it varies with the condition of the joint Its origin may be extra articular due to adhesions and fibrosis of periarticular tissue It is worse at the end of the day after the limb has been in use for many hours and is usually relieved by rest

Or, the pain is intra articular, produced by impingement of soft tissue, this is seen especially in osteoarthritis of the knee joint The pain is definitely brought about by activity and is relieved by rest It also has characteristic sharpness and is momentary and fleeting The pain may also be due to the irritation of raw surfaces grinding together, as one sees for instance in malum coxae senilis or in chondromalacia of the patella Here the synovial reaction is the cause of the pain which is usually relieved by rest Or, the pain may be due to hyperemia in the sclerosed bone, similar to that in Paget's disease This pain is constant, boring and osteocopic It is not affected by rest or inactivity and is rather increased in warm weather The pain may also be due to acute recrudescence of the inflammatory condition, in this case the joint becomes eminently sensitive to exercise and strain This pain is definitely made worse by activity, and it is not always completely relieved by rest Night pain frequently persists

Finally, osteoarthritic pain may not be due to the condition of the joint at

true ankylosis does not occur Even if the joint space has been reduced to a minimum, the articular constituents are still separated

D THE REACTION OF THE PERIOSTEUM

We have already stated that the marginal osteophytes are of cartilaginous and not of periosteal origin In the neuropathic joint, in contrast, the periosteum takes part in the bone formation, and osteophytes can also be found at the insertions of the ligaments and tendons Often the osteophytes will break off and remain in the joint as free bodies

E THE REPAIR OF CARTILAGE

The healing of a cartilage depends on the extent of its degeneration and attrition If the full thickness of the cartilage is destroyed, the bone is laid bare, it then undergoes the already mentioned eburnation with the formation of polishing surfaces On the other hand, if cartilage erosions are shallow, they may fill out with connective tissue, and fibrocartilage may be produced

F IN SUMMARY

The essential pathological changes found in the hypertrophic arthritis are as follows The first stage is the fibrillation and exfoliation of the cartilage, resulting in softening of the cartilage with indentation and depression Then follows the reaction of the bone marrow, which becomes fibrous and forms subchondral cysts or results in the sclerosis of bone where the cartilage has worn away Its most characteristic feature is the renewal of the enchondral ossification at the margin of the joint The bone marrow tongues penetrate into the cartilage, leading to the formation of marginal osteophytes Finally, osteophytes and exostoses may break off and form free bodies in the joint

IV THE CLINICAL PATHOLOGY

A THE GENERAL SYMPTOMS



Fig 49 Hypertrophic arthritis Heberden's nodes at distal and proximal interphalangeal joints Case M S

In osteoarthritis all symptoms develop slowly and insidiously, much more so than in the atrophic type One of the first complaints is stiffness of the fingers, particularly of the distal phalangeal joints The joints become sensitive to weather changes, dampness and rain, there is paresthesia in form of tingling and numbness, although not as marked as in the atrophic type At this time also there appear enlargements of the end phalanges, called Heberden's

motion However, extreme extension and flexion are usually accompanied by an accentuation of these auditory signs, in keeping with the mechanical difficulties which arise at these end points of the arc of motion If the osteoarthritis involves the patella in the so called chondromalacia of the patella, one will find on moving the bone up and down a soft grating, palpable and sometimes audible, as the roughened patella glides over the anterior surface of the femur Loose bodies in the joint manifest themselves by audible clicks, but they are not actually palpable unless they are more superficially situated Sometimes they can be detected as they lie in the suprapatellar pouch

5 Lipoma arborescens

A special type of osteoarthritic knee is the so called lipoma arborescens of Hoffa's disease This is a true osteoarthritis and shows the same histological changes in the bone and cartilage, but it is associated with excessive hypertrophy of the synovial membrane and the fatpads It occurs mostly in women during or after the menopause The motion of the joint becomes more and more restricted, especially the full extension of the knee joint is inhibited There is also considerable secondary muscular atrophy, particularly of the quadriceps, the knee then becomes unstable In addition, the distention of the joint due to expansion of its contents causes a gradual relaxation of the periarticular ligaments Strains and sprains occur regularly, and the distended internal collateral ligament is tender to pressure

As the cartilage becomes thinned out and eroded at the weight bearing areas, a disalignment of the knee joint develops in the form of a genu varum deformity The constant static stresses contribute further to the progress of the arthritis

B THE OSTEOARTHRITIS OF THE HIP JOINT OR THE MALUM COXAE SENILIS

This is another well defined clinical entity of osteoarthritis It involves the hip joint and is seen mostly in patients between the ages of 40 and 60 In contrast to the osteoarthritis of the knee, it is much more frequent in males than in females In two thirds of the cases the symptoms are unilateral at the time of examination, but many times closer observation and the study of the x ray pictures will show beginning changes in the other hip

Clinically the osteoarthritis of the hip presents itself in different stages of development

1 There is an early stage in which the femoral head shows some subchondral cysts and some marginal sclerosis, but otherwise it retains its form

2 Later, hypertrophic changes are seen in the form of marginal exostoses and sclerosis of the head of the femur and wearing off of the joint surface, and the contours show some deformation (Fig 50)

3 Finally, there are advanced cases with prominent symptoms of long standing, signs of marked deformation and subluxation of the head, and very

all, but to irritation of neighboring sensory nerves A good example of this is the radiating pain in osteoarthritis of the shoulder joint, in which radiation extends down the arm and up the neck, or the pain in malum coxae senilis, where the obturator and sciatic nerves are involved in their respective fields of radiation This pain is sharp, neuralgic, and overflowing

d JOINT SWELLING AND ENLARGEMENT

In certain joints enlargement can be noted in more or less circumscribed areas, for instance in the knee joint on both sides of the patella This swelling is due to secondary hypertrophy of the synovial membrane or hypertrophy of the fatpads Some of the enlargement of the joint may be only apparent because of the shrinkage of the surrounding muscles As a portion of the synovial membrane or fatpad becomes impinged, for instance in the knee joint or in the radiohumeral articulation, swelling and tenderness appear in more circumscribed areas representing the sites of impingement The sensitiveness of the hyperemic synovia can best be ascertained by following the outlines of the synovial reflection

V SPECIFIC CLINICAL TYPES OF OSTEOARTHRITIS OF THE EXTREMITIES

A OSTEOARTHRITIS OF THE KNEE JOINT

1 Swelling

It is caused by hypertrophied fatpads and synovial thickening, enlargement of fatpads is noted by the bulging at both sides of the patella The massive synovial hypertrophy causes a fullness at the suprapatellar pouch

2 Tenderness to pressure

This follows the lines of the synovial reflection over the condyles of tibia, femur, and along the suprapatellar pouch Furthermore, with the distention of the capsular apparatus, the posterior capsule becomes tender in the popliteal space Thirdly, because of the strain on the reinforcing ligaments, the tibial collateral ligament will be found sensitive to pressure

3 The mobility of the knee joint

This is usually preserved to a considerable degree, only the extremes of motion are painful because of the presence of increased solid contents of the knee when the synovial membrane and the fatpad are hypertrophied, or because the knee is filled with fluid

4 Auscultatory signs

The osteoarthritic destruction of the knee joint is represented in auditory form by cracking or grating This is usually universally distributed and can be heard in all quadrants of the joint and throughout the entire range of

clinical symptoms appear, for instance in the hip or the knee In others, as in the substragalar articulation or in the ankle joint, complaints may definitely precede any x ray evidence

A THE HIP JOINT

The first sign is the thinning of the cartilage and narrowing of the joint space Then one notices the subchondral cyst close to the joint line, especially in the head of the femur This is then followed by wavy and irregular contours of the joint line and finally the formation of marginal osteophytes The latter become more marked as the process continues, and spicules of bone often break off from their bases and form free bodies We see in some instances that the formation of osteophytes is so massive that it forces the head of the femur out of the socket, producing a subluxation (Fig 52) There is never a true ankylosis, but the x ray picture may be deceiving and the hip may appear ankylosed when it is only mechanically interlocked



Fig 52 Malum coxae senilis Massive exostose forcing head out of socket Case F N



Fig 53 Osteoarthritis of the knee squaring of the patella Case M D

B THE KNEE JOINT

Here we see fine spurs and osteophytes at the margins of the tibial and femoral condyles and sometimes at the spine of the tibia In the side view, the contour of the patella shows irregularities, they seem to be drawn out into fine points and appear more square or rectangular One should not omit an axial picture of the patella to demonstrate signs of cartilage destruction and erosion which characterize the so called chondromalacia of the patella (Fig 53)

C THE ANKLE JOINT

Osteoarthritic changes in the ankle joint are best seen in the side view where they appear as spurs or points arising from the anterior ledge of the



FIG 50 (Left) Malum coxae senilis Marginal exostosis subchondral sclerosis and wearing off of joint surface deformation of the head Case C S
FIG 51 (Right) Malum coxae senilis Massive exostoses narrowed joint space deformed head Case W W

large marginal exostoses The head appears mushroomed (Fig 51), and there are osteoarthritic changes in the acetabulum

THE SYMPTOMS

In the earliest stages the principal complaint is pain on weight bearing with increasing stiffness of the hip Later, the pain becomes more persistent, it may occur at night or even while the patient rests in bed It may assume an excruciating intensity

The outstanding objective symptom is a characteristic position of the hip joint, namely fixation in outward rotation, flexion and adduction Movement in the hip joint is characteristically restricted in the direction of abduction and inward rotation This restriction of inward rotation especially, is one of the earlier objective signs in osteoarthritis of the hip and often precedes the x ray evidence

Due to the outward rotation and adduction contracture the patient has a peculiar gait which is not unlike the gluteus medius gait, because he must swing the body to the other side in order to clear the adducted hip from the ground

VI THE X RAY DIAGNOSIS OF HYPERTROPHIC ARTHRITIS

In contrast to atrophic arthritis, definite x ray findings are often seen in degenerative arthritis even before clinical symptoms appear However, there is no correlation between the x ray findings and the clinical manifestations In some articulations marked roentgenological changes may be seen before the

G THE JOINTS OF THE FINGERS AND THE METACARPOPHALANGEAL ARTICULATIONS

These show arthritic changes early, particularly in the end phalangeal joints The joint fissure appears narrow, the bases of the phalanges are drawn out with pointed ends, and irregularity of the contours is seen Sometimes subchondral cysts are noticed in the heads of the metacarpals (Fig 57)

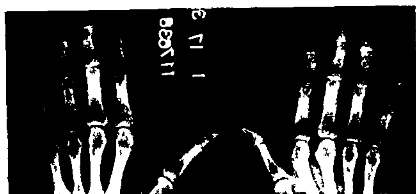


FIG 57 Osteoarthritis of the end phalangeal joint Irregularity of the contours, drawn out bases of the end phalanges Case C S

VII LABORATORY TESTS

Laboratory tests add little to the diagnosis The sedimentation time, blood chemistry and blood count are all normal We have never found any rise in blood calcium, phosphorus or phosphatase, or any change in glucose tolerance The uric acid content of the blood is normal, in contrast to arthritis urica In the synovial fluid the cell count is normal except for an occasional slight increase, which is of little significance

VIII THE TREATMENT

A GENERAL TREATMENT

1 Elimination of focal infection

One focus which is found more often than any other is the diseased tooth In several cases under our observation tooth extraction was followed by striking relief However, such cases are the exception

2 Chemotherapy

So far as the sulfonamides and penicillin are concerned, it is the general opinion that they have no effect whatsoever on the course of hypertrophic arthritis

3 The medicinal treatment

Sedatives and anodynes are given in the more painful stages, otherwise, little can be added in regard to medicinal therapy of hypertrophic arthritis

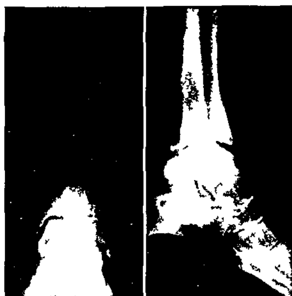


Fig 54 Osteoarthritis of the ankle joint Note the irregularity of the joint contours and drawn out anterior ledge of the tibia Case H B

tibia Corresponding exostoses may be seen at the body and neck of the astragalus (Fig 54) In the subastragalar joint osteoarthritic changes can best be seen in the form of irregularities and blurring of the contours

D THE SHOULDER JOINT

The earlier signs are narrowing of the joint margin, subchondral cysts in the humeral head, and slight elevation of the head in the glenoid fossa Exostoses usually appear later, and they are not infrequently seen at the margin of the glenoid

E THE ELBOW JOINT

The first signs of osteoarthritic changes are usually seen in the radiohumeral articulation and particularly at the head of the radius (Fig 55)



Fig 55 (left) Osteoarthritis of the elbow joint with free bodies Case M T
Fig 56 (right) Osteoarthritis of the wrist Note irregularities of the contours and sclerosis in the radiocarpal articulation Case G W

F THE WRIST JOINT

Here we must look for the contours between the proximal carpal row and the radius for the first indication of degenerative changes (Fig 56)

impingement, may also yield to manipulation, but the principal feature of an enlarged fatpad is mechanical in effect, and if it is possible to decrease its size the impingement may disappear If such is impossible, only removal of the fatpad will remedy the situation

Pain due to the grinding together of roughened articular surfaces and sub sequent synovial irritation calls for elimination of weight bearing or cessation of motion Tolerance to weight bearing must be developed gradually If the joint remains persistently intolerant, both to weight bearing and motion, one may have to resort to operative measures Fusion may be indicated

Again, if the pain is due to acute inflammation and reaction of the synovial membrane, the indication is rest, massage and heat, and eventually x ray therapy for the more acute episodes The joint must be immobilized or at least kept at rest during the acute phases

We see, therefore, that the local treatment is, first, rest, then physiotherapy, both at the proper time To a large extent these measures will be able to control pain and swelling and thereby automatically increase the range of motion It is important that under this treatment the supporting and protecting musculature be developed properly, because it contributes greatly to the stability of the joint

As soon as immobilization and rest are no longer necessary, one should proceed to heat and massage treatment and the application of graded exercises, as dictated by the experience and common sense of the physician Warm baths, therapeutic pools, and hand baths come into practical use in these situations In general, it makes little difference what kind of heat is applied, but one must be sure that the intensity of the heat furnished by the selected modality is tolerated If a joint is acutely irritated or inflamed, it will not stand deep and penetrating types of heat In the same sense, one must undertake graded exercises with caution Any discomfort which follows such exercises would indicate that the safety limits have been overstepped

2 Is manipulation of arthritic joints permissible?

For contractures due to spasm of muscles or to shrinkage of periarticular structures, manipulative treatment is justified under special precautions Manipulative treatment in cases of intra articular adhesions is *never* justified The condition is somewhat different in the hypertrophic arthritis because no true intra articular adhesions exist In the malum coxae senilis it could be decided according to the type of pain, whether it is intra articular due to the grinding and the synovial reaction to it, or whether it is extra articular due to capsular shrinkage or muscle contracture We can readily see that in the latter case careful manipulation may be justified

3 Shall braces be used as supports for weight bearing joints?

We have already referred to this point in discussing atrophic arthritis Any complaint which is referable to the periarticular reinforcing ligaments calls

Certainly salicylates have no effect or at most, only a temporary one Lately physostigmine has been tried [4] This drug relieves spasm and pain in a short time, and is given in doses of 0 6 mg in connection with atropin sulfate In cases of fibrositis or bursitis, as well as arthritis, good results have been reported

4 The hygienic and dietary treatment

There is no special diet The only restriction is a reducing regime in obese patients, especially in cases of osteoarthritis of the knees with fatpad enlargement

We usually allow 1000 to 1500 calories for weight reduction, but one should be warned that a rapid weight loss is not advisable in patients in the age group of this type of arthritis Not infrequently the metabolic rate is low In that event, we give 0 5 grain of desiccated thyroid It is obvious that starchy foods, pastry, sweets, heavy meat, cocoa and chocolate should be avoided as much as possible There should be ample fluid intake

5 The vitamins

In osteoarthritis, the administration of vitamins does not have the importance that it has in rheumatoid arthritis, because the patient is not deficient in vitamins to the same degree Nevertheless, it is well to furnish adequate vitamin B_1 or thiamine chloride in doses of 1 to 5 mg two to three times a day or, still better, in daily doses of 50 mg intravenously or subcutaneously

So far as vitamin D is concerned, it has no particular specific value, except that it contributes to improving the general health of the patient It must not be forgotten, however, that many patients with senile osteoporosis will show secondary arthritic changes, especially in the spine In these cases there is a definite vitamin D deficiency with inadequacy of calcium intake, which may have existed for many years, and vitamin D therapy is essential

B THE LOCAL CONSERVATIVE TREATMENT OF OSTEOARTHRITIS

1 Control of pain

Analyzing the Indication

The principal indication for the local treatment is pain From what we have said about evaluating the different types of pain, it is obvious that one must obtain a good description so as to determine the direct source of the pain sensation If the pain is osteocopic and associated with tension in the bone, one must decide on some alterative and counter irritant method It is here particularly that deep x ray therapy or even drilling of the bone will give relief On the other hand, if the pain is due to adhesions or to extra articular fibrosis, the treatment may be manipulation under anesthesia

Pain which is due to impingement of soft tissues, as in the case of fatpad

roughness of the joint surfaces with marginal exostoses interfering with the function of the joint 3) In cases of synovial impingements which do not react to rest or physiotherapy and which cause repeated episodes of joint irritation 4) In cases of extensive and irreparable destruction of the joint surfaces which have passed any possibility of spontaneous regeneration

1) (Group 1) Patellar shaving and patellectomy This group involves cases of roughness and irregularity of the patella with fibrillation of the cartilage sufficiently extensive to constitute a constant source of irritation Here the removal of the patella is indicated This operation has been used for many years in traumatic conditions, especially in comminuted fractures As an operative method to relieve or improve conditions in hypertrophic arthritis, however, it is of more recent date Haggart[7] reported 20 cases of degenerative arthritis of the knee joint treated by removal of the patella as well as the synovial tags, of these, 19 showed improvement The rationale of removal of the patella is the relief of mechanical friction The operation is specifically indicated in advanced cases of chondromalacia The indication for complete removal of the patella cannot always be made before the operation It depends upon the degree of fibrillation whether simple removal of the affected cartilage will suffice or whether the patella will have to be excised If the fibrillation has not reached too great a depth and if there is still a sufficient layer of healthy cartilage covering the bone, shaving the patella may be sufficient, a procedure which we will discuss later in connection with the so called Magnuson method If it appears that the degeneration has reached the full thickness of the cartilage and has occupied large plaques of the cartilaginous surface, it is safer to remove the patella

The question has often been brought up what removal of the patella means to the mechanics of the joint Some authors have gone so far as to maintain that the removal of the patella has no mechanical influence upon the knee joint whatsoever Such a statement is, of course, not justified by the principles of mechanics It is quite correct to say that the removal of the patella does not essentially interfere with the function of the knee joint, but this does not mean that the removal has no mechanical effect whatsoever upon the knee There is a close relationship between the patella and the quadriceps, since the patella is a sesamoid bone implanted into muscle Its mechanical function is, however, of comparatively less import, so that in a given case the disadvantage of removing the patella may be far outweighed by the advantage of relieving the pressure and friction which causes increasing osteoarthritic changes and disability of the joint

Our statistics on the removal of the patella for chondromalacia (H Brindley) are as follows From 1925 to 1948, 18 patellae were removed from 16 patients, ranging in age from 15 to 63 years The duration of symptoms was from three months to 15 years Results were good in 10 patients, or 60 per cent, with stable joints, no pain or swelling, and walking without difficulty In three patients the results were fair, in two patients, in whom both patellae

for support by mechanical means This may be accomplished by bandages, especially in the knee

There are cases of osteoarthritis of the knee in which, in the course of time, the ligamentous relaxation has progressed, the joint is lax and becomes dis aligned One can easily see that in a case of this type braces become very essential as a means of support They are, however, only temporary measures and can be abandoned as soon as recovery of the overstretched ligament has taken place and the muscles have developed to the degree where they can provide adequate support to the affected joint

Case L C (Male)	#54 864
Age 62 years	Adm October, 1944

This patient had trouble with his left knee for the last 30 years At the time of admission he had a very painful knee with a range of motion of only 5 degrees following an injury five months previously The x ray revealed marked osteoarthritic changes This patient was advised to have the knee fused, but he rejected the operation and preferred to wear a long leg brace He worked constantly, and in doing his farm work had experienced no pain at all, although he had not bent his knees for two years when seen last At that time the examination showed little if any motion in the knee joint, and there was no pain on walking or standing either with or without the brace An x ray was taken three years after first admission and showed the same osteoarthritic changes with a markedly narrowed joint space

This is an example of osteoarthritis of the left knee in a 62 year old farmer who wears a brace for relief of pain and is thus enabled to do heavy work Evidently the knee joint took care of itself, and the last x ray picture showed what appeared to be an obliteration of the joint space The knee had become entirely painless so that he was finally able to discard the brace

Contracture of the toes and particularly the metatarsophalangeal articula tions should be prevented by proper support for the arches and by manipula-tion when actual contracture has developed The conservative measures to be applied after manipulation are the same as those used in static flatfoot or static clawfoot

C THE ORTHOPEDIC TREATMENT OF SPECIFIC CLINICAL TYPES OF OSTEOARTHRITIS

1 Hypertrophic arthritis of the knee joint

The indications for *conservative treatment* have been discussed under the heading of local conservative treatment of osteoarthritis

The Operative Indications

Operative procedures should be carried out in the following groups of cases

1) In cases of roughness and fibrillation of the patellar cartilage with in creasing osteoarthritic reaction in the joint 2) In cases of irregularity and

called Lipoma arborescens or Hoffa's disease In degenerative arthritis this impingement is secondary to primary cartilaginous changes Removal of the synovial membrane does not abolish an active focus of infection as was formerly believed It is simply a procedure to remove a mechanical obstacle In the majority of cases one will find that after removal of the fatpad the knee can be completely aligned and straightened out

In cases in which the cartilage is severely damaged we recommend combining the Magnuson operation with removal of the fatpad or, if necessary, with removal of the entire synovial membrane The latter procedure, however, is not often indicated in the degenerative type of arthritis

4) (Group 4) The fusion operation This procedure is indicated in Group 4 in which the destruction of the joint is extensive and irreparable and where there is no probability of a spontaneous repair A great deal of judgment is required to determine whether or not the motion of the arthritic knee should be completely sacrificed In most instances the decision cannot be made until the joint is exposed at the operation If the joint appears hopelessly destroyed its elimination is fully justified, as a fused knee holds out the promise of a painless, weight bearing leg

One important point is to select the position in which the knee joint is to be fused We emphasize that it should never be fused in full extension, except in cases of considerable shortening The favored position is flexion of 15 to 25 degrees, depending upon the occupation of the patient Those having a standing occupation, such as farmers and artisans, require less flexion Those having a sedentary occupation should have at least 25 degrees of flexion Our technique consists of removing the cartilage of the tibia and femur completely and then implanting the denuded patella as a graft into grooves prepared in the femur and tibia The grooves should be so designed that when the patella is firmly implanted the knee is in the desired degree of flexion, preferably of 25 degrees This is essentially the technique recommended by Hibbs

5) Arthroplasty of the knee in hypertrophic arthritis We have already observed that in general the results of arthroplasty in arthritis are none too good, and they are worse in the knee than in any other joint Our results of arthoplasty of the elbow, hip and knee in hypertrophic arthritis were 70 per cent good, 20 per cent fair, and 10 per cent poor The over all good results in the knee joint, however, were only 39 2 per cent [3]

One should not undertake an arthoplasty of the knee until one is sure that the joint is completely quiescent, and the operation should not be considered if the knee is well fused in good position and is painless The arthroplasty is justified if both knees are ankylosed, and in one knee if ankylosed in unsuitable position The security which the completely stiffened knee gives to the gait of the patient, provided it is in suitable position, permits one to chance an arthroplasty on the other side

were removed, the result was good in one and poor in the other In one case, reformation of the patella occurred and it had to be removed

2) (Group 2) The Magnuson operation This operation is indicated in cases belonging to Group 2, where joint irregularities and roughness of the marginal contours are more extensive Although degeneration of the joint cartilage is moderate, it extends over a large portion of the joint surface, and there are numerous marginal exostoses which interfere with free motion

The operation consists in the removal of all mechanical obstacles, especially the exostoses and lippings and, in addition, in the shaving off of the diseased cartilage Impinging fringes of fatpads are also removed The method is par ticularly useful in cases with osteophytes and hypertrophied fringes, all of which produce mechanical obstructions

The question is, how much cartilage one may shave off without actually interfering with the mobility of the joint, in other words, how much cartilage can be removed without producing intra articular adhesions It is obvious that only degenerated portions of the cartilage should be removed so that the rest can form a satisfactory gliding surface of the joint from the new fibrocartilage The cartilage which has been broken away, so to speak, from its mooring to the subcortical bone has no tendency to fill in from the side, since cartilage grows in straight columns from the matrix to the surface It does not grow side wavs as does the epithelium Consequently, if an area of cartilage is gouged out, including the matrix of the cartilage, then this cup shaped area of defect has no tendency to regenerate, but it fills with fibrous tissue which comes from the bone marrow It follows therefore that the operation can be carried out only so long as the degeneration does not include the matrix of the cartilage It is not necessary in these cases to remove the entire synovial membrane, but only as much as constitutes the mechanical obstacle The operation must be followed by very careful and prompt after treatment Motion is started as early as the fourth day Eight or 10 days later the patient is allowed up on crutches, and soon thereafter careful weight bearing attempts can be made

Magnuson[15] reports 62 cases of degenerative arthritis of the knee operated on by this method, 60 of which made a complete recovery during observations ranging from six months to 16 years G E Haggart[8] recently reported 30 cases, 27 of which gave good results with increase of motion, the minimum range being 67 degrees, in 21 cases the motion returned to normal We per formed 10 Magnuson operations for osteoarthritis of the knee between 1942 and 1945 The results in our 10 cases were excellent in four, good in two, and poor in four (Hall[9])

3) (Group 3) Synovectomy in degenerative arthritis This operation is indicated in Group 3 comprising cases of synovial impingement which do not respond to conservative means Synovectomy consists in the removal of the synovial membrane when the latter forms a mechanical obstacle It is therefore especially indicated in cases of impingement, particularly in the so

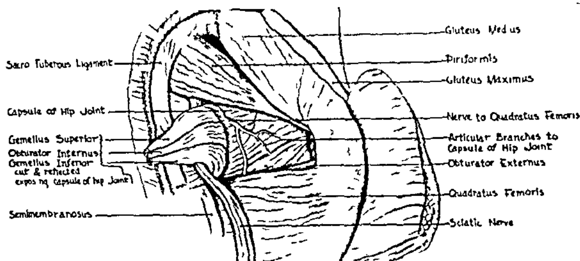


Fig 59 Sciatic nerve supply of the hip joint (Lowry and Ergenbright[1])

bations of pains are best treated by rest and traction, muscle spasm is best taken care of by plaster cast and crutches, as is also the intra articular pain In older individuals, particularly if obese, in whom it is not practical to apply a cast, one may try crutches alone, with the support of physiotherapy

Procaine injection In some cases the pain is not relieved by traction, or it returns after a very short period of remission Temporary relief can be obtained by procainization of the sensory nerve supply of the hip joint[14] The nerve supply to this joint comes from the obturator and the sciatic nerve The obturator nerve (Fig 58) sends fine twigs from its deep branch into the medial and anterior portions of the capsule of the hip joint These twigs branch off very close to the obturator foramen The posterior supply of the joint comes from a fine branch of the sciatic nerve This branch splits off from the anterior surface of the trunk and supplies the posterior portion of the hip joint It can be located at the lower border to the piriformis muscle (Fig 59) If one constructs a line between the posterior superior spine and the sacrococcygeal junction and divides the distance in three equal parts, the nerve will be found lying along a line drawn from the junction point of middle and lower thirds to the greater trochanter (Fig 60)

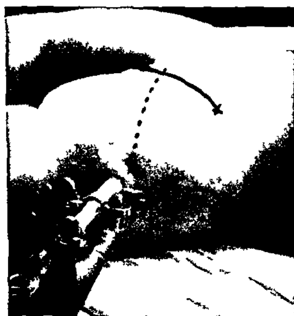


Fig 60 Locating sciatic branch to hip joint on a line extending between the tip of the greater trochanter and a point two thirds the distance between the posterior superior spine and the sacrococcygeal junction

Both nerve supplies can thus be located easily and a 1 per cent procaine solution injected into the re-

2 Malum coxae senilis

a CONSERVATIVE TREATMENT REST, IMMOBILIZATION AND PHYSIOTHERAPY

A good deal can be accomplished in the early stages of hypertrophic arthritis of the hip by conservative measures which do not necessarily interfere with the patient's occupation At this stage, the patient complains of increasing stiffness of the hip joint, particularly in the morning, and of pain when he walks long distances However, he is still free from night pain, and the examination will show a fair range of motion in extension and flexion, and possibly even some in abduction and adduction, although there is a definite restriction in inward rotation It is very important to start treatment at this early stage It should consist first in the elimination of the infectious focus and in the restoration of the general health and resistance of the patient Of great advantage is a short period of bed rest combined with traction Under this regime the pain is usually controlled within a few weeks This treatment is particularly appropriate at the time night pain begins to appear It can well be combined with physiotherapeutic measures, such as application of heat, light massage, and the development of the abductor and extensor muscles of the hip In cases in which traction relieves the pain in recumbency, but in which pain persists on ambulation, we find it best to apply a plaster cast and put the patient on crutches The cast is left on for six weeks, and it is more suitable on the whole than a tuber seat brace, which is quite uncomfortable in obese patients Usually the type of pain gives some indication to what extent recumbency and traction will offer relief Night pain, particularly if spasmodic, is due to irritation of the synovial membrane Pain on walking is mostly due to the pressure and grinding of the bone surfaces against each other More indefinite pain developing during the day and increasing evenings is usually due to strain of the surrounding musculature It would seem, therefore, that periodic exacer

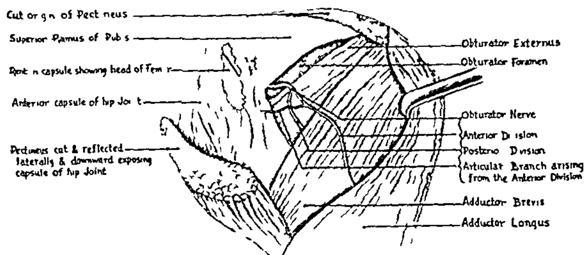


FIG 58 Obturator nerve supply of the hip joint (Lowry and Ergenbright)

(1937) reported 108 cases, stating that he obtained improvement in 80 per cent He drilled holes through the head and neck of the femur in order to obtain a decompression effect Henderson,[11] applying the method in the early stages of the disease while there was still rarefaction of the femoral head and of the superior border of the acetabulum, obtained initial improvement but found that the relief of pain was not lasting Tavernier[20] recently reported 12 cases with immediate relief of both spontaneous and elicited pain It seems, however, that the later results are not as good as the immediate ones While his earlier results were excellent, checkups five and seven years after the operation revealed that in nine of his 12 cases the pain returned in intervals varying from two months to two years



Fig 62 Fusion of the hip joint for osteoarthritis combined intra and extra articular method

The drilling is done through the neck and head of the femur from the base of the greater trochanter, at an angle of 120 degrees, in the direction of the neck The position can be controlled by x ray The effect this operation has on the control of pain is not quite clear The relief is evidently due to the changes which are caused in the vascularization of the affected bone, and is in line with the old idea of Ollier, who trephined painful hyperostoses to control pain

Method of revascularization of head and neck by muscle implantation for the control of pain The underlying idea is to secure a better nutrition for the femoral head by establishing an additional circulatory supply from muscles implanted into the neck Venable and Stuck[21] mention that this method was first suggested by Brainerd 90 years ago The authors experimented for several years with the increase of blood supply in the hip by this muscle flap transplantation They used the vastus lateralis which they mobilized and then transplanted a flap into a long trough made in the neck of the femur This is a most original and interesting idea which deserves attention, even though there are at present no definite operative end effects to report

Fusion operation for the control of pain Many surgeons believe the fusion to be the operation of choice and the only one which will definitely dispose of the symptom of pain Most certainly it is the safest method of eliminating pain as well as of restoring a suitable position of the joint In many cases it is difficult to decide in favor of this radical measure because the patient still has a considerable range of motion or his pain is only intermittent Or he may not be a proper risk for the operation and the long period of recumbency and immobilization following it

Persistent pain and particularly night pain with a rapidly decreasing range

gion We have carried out this procedure in a number of cases and found that the relief lasts from several days to several months, the injection can well be repeated after a suitable interval (Fig 61)

All injections were done with 1 per cent procaine, 10 to 15 cc each being used for the anterior and posterior regions The results of these injections were tabulated by Lowrie and Ergenbright[14] as follows Of 23 patients treated, 19 had immediate and complete relief of pain both on rest and on motion, and four cases had little or no relief



Fig 61 Malum coxae senilis procaine injection Case E S (male) 49 years December 1946 Progressive pain and gradual decrease of motion in the right hip for 15 years X ray shows osteoarthritis of the right hip Procaine injection into the obturator and sciatic supply of the right hip brought complete relief of pain and ability to walk without support No recurrence of pain for three months second injection also resulted in relief of pain

The x ray treatment of osteoarthritis of the hip In addition to the treatment by recumbency or traction, certain cases of intense and localized hip pain, particularly those with persisting night pain, are benefited by x ray treatment Our routine is to subject the patient first to traction, immobilization and physiotherapy, then to procainization and then, failing in these, to x ray treatment

This treatment was carried out in seven cases (Lichtor), each course consisted in 13 treatments of 100 R at 48 hour intervals A total of 14 x ray courses was given to all seven cases Eight of these courses were followed by marked relief of pain within several days, in two more, relief was obtained within a period of one week or more, in the four remaining courses of x ray radiation, there was no relief Even in the cases which were relieved, the pain returned within three months Our conclusion was that the x ray treatment, while giving temporary relief in many cases, is not of lasting value in the treatment of malum coxae senilis

b Operative Treatment

The operative treatment pursues three objectives The control of pain, the principal objective, the correction of the deformity and, thirdly, the restoration of mobility of the joint

1) Operative methods for the control of pain Method of bone drilling The originator of the drilling of bone is Arnaud, who first mentioned the method in 1928 He intended it less for the control of pain than for the control of the arthritis itself, apparently taking his cue from the concept of Robertson Lavalle (1919) who advocated the method of forage or drilling in the treatment of tuberculous arthritis, particularly of the knee joint Four years after Arnaud, Duvernay (1932) reported 10 cases of osteoarthritis of the hip in which the pain had been relieved by drilling of the neck MacKenzie

placing it in a plaster cast gives bony union in only 60 per cent of the cases, and that the functional results are poor

Favorable results from arthrodesis in hypertrophic arthritis of the hip are also reported by Ghormley[6]

The nerve resection for relief of pain This is a method exclusively designed to relieve pain in hips in which the arthrodesis is either rejected or is inadvisable because there still remains a large excursion field of the hip joint, or in cases of bilateral involvement where one hip must necessarily be left in a movable condition The procaine injection, we have mentioned above under the heading of conservative treatment, should precede the nerve resection, since it gives information regarding the origin of pain and the chances for its suppression by eliminating the sensory nerve supply of the joint If the pain is completely relieved, even for a short time after procaine injection into the fields of both the obturator and sciatic nerve supply, one may then proceed to resect the sensory branches in order to establish a more permanent state of relief The method was first mentioned by Camitz of Gotenburg, Sweden (1933), and was later taken up by Tavernier,[9] who described an operative plan of dissecting the deep branch of the obturator nerve in order to suppress the pain

The first report published by Camitz of his results on resection of the nerve was favorable The pain was suppressed Tavernier and Godinot[9] (1939) reported a series of 57 obturator neurotomies of which 45, or 82 1 per cent, gave immediate relief The results, however, were not lasting, and in their later report of 51 cases they could find only 14 in which the resection was completely successful, making the total of satisfactory results 38 per cent The cause of the failure was then recognized in the fact that after denervation of the obturator nerve supply, pain still remained in the posterior territory This led them to complete the procedure by adding the denervation of the posterior nerve supply to the hip joint capsule

The combined method of anterior and posterior resection is now used by Tavernier and Godinot[9] as a routine, and the results have been much more satisfactory The patient is immediately relieved of pain, and after about 10 days he can begin to walk again Of their 24 cases operated on, 17 or 70 per cent were completely relieved immediately after the operation Inasmuch as the combined operation is new, having been performed for the last few years only, there are as yet no authentic reports on the late end results available

Our own statistics (R W Newman) on nine nerve resections are as follows In four cases in which the obturator branch alone was resected, three were relieved of pain, of five cases in which both obturator and sciatic supplies were resected, four were definitely relieved and one case was improved However, the observation time averaged only nine months

The simplicity and effectiveness of articular denervation assures this method a welcome place in the treatment of malum coxae senilis, particularly in cases which do not respond to simple methods of immobilization and physiotherapy, yet are not advanced enough to warrant a radical procedure such as arthrode

of motion and increasing deformity constitute a definite indication for this operation Of all procedures for the control of pain, it is without doubt the most reliable The fusion can be carried out by any one of the numerous technical procedures that have been advocated We prefer the intra- and extra articular types of fusion For the intra articular type we use the Albee technique combined with a bone graft or strut placed in the trochanter and the os ilii (Fig 62) It is not necessary to dislocate the head in order to obtain complete ablation of the cartilaginous covering, especially since this is a rather shocking procedure poorly tolerated by many patients Dickson and Willien[5] advocate a simpler procedure The hip is exposed by a Smith Peterson incision, and the cartilage on both surfaces of the joint is broken up by a special curved chisel such as the one devised for arthroplasty This exposure allows for the correction of the flexion and adduction deformity, as well as for sufficient roughening of the cartilaginous surfaces When the joint has been adequately freed by this method, it is possible to manipulate it without dislocating the femoral head and to give it the proper position, which is flexion of about 30 degrees, abduction of no more than 5 degrees and slight external rotation After this is done, a vitallium nail is driven into the neck and head of the femur through a second incision, and the nail is carried into the thick portion of the ilium above the superior roof of the acetabulum The Dickson and Willien operation has the advantage that there is no operative shock since the hip is not being dislocated, and no application of plaster of Paris is necessary In all 10 cases which these authors reported there were no postoperative complications, and complete fusion of the joint was accomplished

For the extra articular type we use the greater trochanter, according to the methods of Hibbs and Hass Tavernier and Godinot[6] obtained rapid and complete ankylosis in three months with the combined method of intra and extra articular fusion Before such a fusion operation is undertaken, one should ascertain the mobility of the lumbar spine, since the ankylosed hip depends upon the aid of spinal movement for carrying out flexion and extension of the limb If the mobility of the lumbar spine is good, then one can allow more flexion in the hip joint, which makes it easier for the patient to sit In many cases, however, we find that the motion of the spine is considerably restricted due to spondylarthritis In this case, the hip must be given a position of less flexion, possibly not more than 10 to 15 degrees

Niebauer and King[17] use a nail, following the method first reported by Watson Jones[2, 3] They found that immediate fixation could be obtained with the nail alone, but both rigidity and strength of fixation are greatly increased when one inserts a screw through the acetabulum into the head at a right angle to the nail With this operation which offers a very firm fixation of the hip joint, the authors obtained painless, stable hips in 15 of 24 cases the patients were encouraged to sit up in a chair as early as the 10th day Watson Jones[2] also favors the nail arthrodesis for the unilateral osteoarthritis of the hip joint, pointing out that the simple method of denuding the joint of its cartilage and

stresses in walking as well as in standing, one must first make sure that the joint has a sufficient range of motion to allow for the necessary flexion range after the osteotomy is performed, secondly it must be established that at least a portion of the joint surface is weight tolerant

We may state then that the cases suitable for osteotomy are those which have no night pain and do have at least some pain free adduction and flexion range This point is important They must have a painless range of motion of adduction and flexion, because after the osteotomy the flexed limb is extended, and the adducted limb is abducted Consequently, as the limb is flexed from the extended position or is adducted from the abducted position, it must have at its disposal a certain painless flexory and adductory field The only remaining condition then is that the lumbar spine have sufficient mobility left to take care of the forward flexion of the body as the patient sits down The type of osteotomy is not of great consequence One may use either a simple osteotomy or the Schanz technique, or a combination of osteotomy and pinning to secure the position of the fragment

The practice of combining osteotomy with fusion has a certain rationale Since there are no muscles attached to the upper fragment, the osteotomy immobilizes it, provided the osteotomy is performed above the lesser trochanter Consequently, the success of the fusion is more assured

Statistics In our cases of subtrochanteric osteotomy for the correction of position in malum coxae senilis, the results were gratifying Early results of the simple subtrochanteric osteotomy were 73 per cent good and 23 per cent fair, taking in consideration not only the position but also the weight bearing ability of the joint

3) Operative methods for restoration of mobility *Acetabular plasty* Several years ago, Smith Peterson of Boston devised a procedure called the acetabular plasty or cheiloplasty The operation consists in the removal of the overhanging bone ledges and bridges around the acetabulum and around the head of the femur It is indicated in cases where the exostoses around the head are massive, but the joint surfaces themselves are still smooth The field for this procedure is rather narrow The efficacy of it is still unsettled, but it seems to provide an increase in the abduction range

Arthroplasty Of all the operations performed on painful and stiff hip joints due to osteoarthritis, this is the most difficult and the most uncertain in its end result It must be accepted, therefore, only with a great deal of reservation, and the indication must be drawn particularly strictly The individual must be young and vigorous, preferably under 50 years We do not think it suitable for heavy workers, but in bilateral involvement of the hip joint especially when motion has been practically obliterated, it is definitely indicated, because it is most desirable to secure motion in one hip (Fig 63)

Whether the cup arthroplasty or fascial interposition is the better method is still a question The vitallium cup method is certainly simpler Harmon[10] uses a sort of lucite or plexiglass material to cover the head This material is

sis The question may be brought up whether the denervation of the nerve supply of the hip joint involves the danger of producing a neuropathic joint, such as we see in tabes It seems to us that this danger is remote since we see no such joint degeneration in other conditions in which the peripheral nerve supply is interrupted, whereas we do see it in pathological conditions involving the spinal cord itself We believe, however, that a more selective indication is necessary The simplest instance is the case of arthritis with painful hips, which otherwise would have to be arthrodesed because of pain, but which still have a considerable range of motion In bilateral cases of this kind, the indication may be particularly appropriate since it would be unwise to carry out the arthrodesis on both hips One would then have only the choice between arthrodesis of one hip and arthroplasty of the other As we shall see later, the arthroplasty operation of the hip does not give uniformly good results, and it seems to us advisable to proceed with denervation of one hip, if not of both, before such an arthroplasty is performed

Finally, there are numerous patients who are too senile and too frail to tolerate an extensive operation such as arthrodesis or arthroplasty

Section of Obturator and Sciatic Branches

Case A T (Female) #41 8132
Age 66 years Adm July 25, 1947

Osteoarthritis of the right hip with obturator and sciatic radiations Temporary relief from procaine Section of obturator nerve performed in September, 1947, and of sciatic branch in December 1947 End result was complete relief

2) Operative methods for correction of deformity We may assume that one joint has adequate tolerance for weight bearing or has some range of motion, but the faulty position causes difficulties in walking and produces a strain at the lumbar spine This is a case for the osteotomy Before an operation is decided on, one must take thorough stock of the general body alignment to ascertain for instance, if there is increased forward inclination of the pelvis which causes the weight to be borne on the posterior aspect of the acetabulum and the corresponding anterior portion of the femoral head

Only in early stages do conservative means suffice to correct the flexion and adduction deformity of the hip According to Kuhns,[1] correction may be managed by exercises or by cast treatment or traction, according to the particular case If there is muscle spasm, cast or traction will suffice If there is muscle disequilibrium, exercises of the particular muscles are in order To correct the increased forward inclination of the pelvis, back support and postural exercises are helpful If pain is associated with muscle spasm, one will necessarily start first with rest in bed and then proceed with the application of a hip spica and crutches to secure weight bearing tolerance and ambulation

If under this treatment the faulty position of the hip with the attending inclination of the pelvis cannot be corrected, the osteotomy is indicated Since following corrective osteotomy the hip joint must sustain weight bearing

labor, one being able to go back one year after the operation, and the other, after three years The observation time varied between eight months and four years

The Acrylic Prosthesis The latest contribution to the operative treatment of the osteoarthritic hip is the acrylic prosthesis introduced by the Judet brothers[114] in Paris in 1946 It consists in the removal of the head and its substitution by a prosthesis made of acrylic material, or in its later modification of Vitallium or stainless steel This method is arousing an unusual amount of interest not only as a substitution for the arthroplasty, as in malum coxae

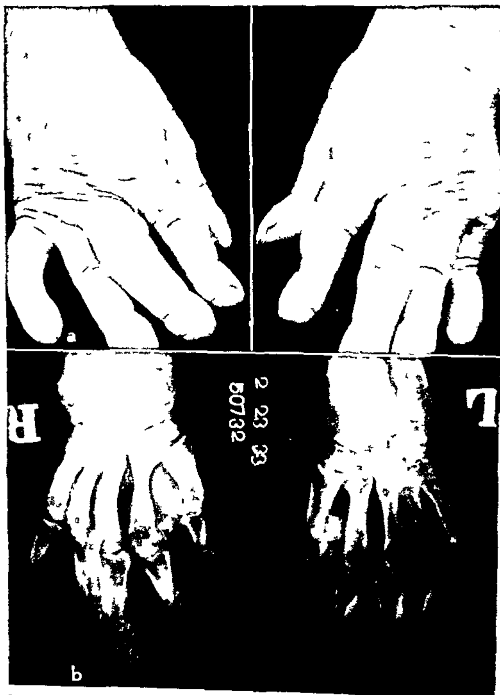


Fig 64 Pulmonary hypertrophic osteoarthropathy Case M W a) Note clubbing of the end phalanges b) X ray shows advanced hypertrophic changes

flexible and durable and has great tensile strength. The after treatment in arthroplasty of the hip joint is traction for several weeks, while the hip is held in extension and inward rotation. Active exercises are begun in one week. Mobilization should be slow, and no force should be applied. So far, any attempts to improve the range of motion by forcible maneuver have failed to give us results. If contractures occur following the arthroplasty, it is a sign the arthritis is still active. The patients are allowed upon crutches six weeks following the arthroplasty. Weight bearing is not permitted, however, before three months. The amount of flexion obtained is usually moderate and rarely surpasses 45 degrees. In cases which have 30 degrees of motion or more before,



Fig. 63. Malum coxae senilis, cup arthroplasty. Case G. F. (female) #42 2122, 32 years. February 1942. Pain and stiffness of the left hip for three years. X ray shows osteoarthritis. Arthroplasty with Smith Peterson cup resulted in a good range of painless motion. a) Before. b) After operation.

the chances for obtaining more extensive ranges are much better. In the series of 16 cases reported by Harmon,[10] 13 of which were observed for 18 months or more, excellent results were obtained in 62.5 per cent. The statistics of Bickel, Ghormley, Coventry and Mussey on 27 cases of cup arthroplasty gave 15 per cent excellent, 30 per cent good, 27 per cent fair, and 30 per cent poor results. The results of Tavernier[9] are somewhat better. Of 17 cases, he reported 12 excellent results with a mobility of 30 to 40 degrees flexion extension range, and 15 to 30 degrees abduction. This standard, however, is lower than that used by American authors. Statistics on our results[4] comprise 13 cases operated up to 1939. Of these, seven or 54 per cent gave good results, two or 15 per cent, fair results, and four or 31 per cent gave poor results. The cases with good results had at least 30 degrees of flexion range with 10 degrees of abduction, and were painless on weight bearing. Those with fair results had a flexion range of less than 30 but more than 20 degrees, no adduction contracture and no or only slight pain on weight bearing. It was found, however, that there were only two patients, or 15 per cent, who could really do heavy

11 HENDERSON M S and POLLOCK, G A *J Bone & Joint Surg*, *12* 923 1940
11a JUDET, J Utilization of the acrylic prosthesis Acta chir Belg *6* 550 (July Sept) 1950
12 KUHNS J G *J Bone & Joint Surg* *24* 547 1942
13 LANGE, F J *Handb d Spec Pathol Anat u Histolog* Berlin O Lubarsch and F Henke 1939 *92* *252* 1934
14 LOWRY F C and ERCENBRIGHT W V Personal Communication
15 MAGNUSON P *Surg Gynec & Obst* *73* 1 1941
16 NICHOLS E H and RICHARDSON F L *J Med Res*, *21* 149 1909
17 NIEBAUER J J and KING D *J Bone & Joint Surg* *28* 103 1946
18 POMMER G *Denkschr d k Akad d Wissensch* *89* 65 1914
19 TAVERNIER L C *Lyon chir* *36* 228 1939
20 TAVERNIER L C and GODINOT C H *The Surgical Treatment of Malum Coxae Senilis* Paris Masson & Co 1945
21 VENABLE V S and STUCK W G *Ann Surg* *123* 641 1946
22 WATSON JONES R *J A M A* *10* 278 1938
23 ——— *Proc Roy Soc Med* *38* 7 1945
24 WRAY R M *Research Seminar Notes,* Dept Orthop Surg State Univ of Iowa *12 D* 65 1938

senilis but also because it has many other fields of application The Judet brothers report to date on no less than 500 cases, Dr J Verbrugge of Antwerp on 300, and they are joined by many American surgeons, W R McAusland of Boston being one of the most enthusiastic supporters of the method The acrylic material is being supplanted gradually by metallic material (Vitallium or stainless steel) in a number of modifications (Jaennichen, Peterson, Eicher and so forth) So far the reports are extremely optimistic, though late end results cannot yet be reported Particularly the mechanical effect of the prosthesis upon the weight bearing tolerance and the stress reaction of the supporting neck and shaft of the femur is yet to be determined

IX ON HYPERTROPHIC OSTEOARTHROPATHY

This is a condition resembling hypertrophic arthritis which develops in the course of severe circulatory disturbances particularly chronic diseases of the lungs, such as lung abscess and bronchiectasis It was first described by Bamberger The clinical picture consists essentially in the stubbing of endphalanges of the fingers and toes, and a crinkling and furrowing of the nail The x-ray shows osteoarthritic changes in connection with secondary periosteal thickening of the bone, particularly of the phalanges and metacarpals

Case M W (Male) #38 30059
Age 65 years Adm May, 1939

In January, 1930 the patient began to complain of marked enlargement of the hands, wrists forearms, feet, ankles and the legs At the age of 15 he had measles followed by an enlargement of the joints and the 3rd and 4th fingers on the right hand Every spring since, he had recurring swelling with pain of the feet and hands, each attack left the members enlarged Patient had been unable to walk for two years or to work with his hands for three to four years Examination revealed enlargement of the forearms, wrists hands, legs ankles, and feet with marked arthritic changes The electrocardiogram revealed a bundle branch block, and the lungs showed chronic bronchitis The fingers had the characteristic stubbing of the endphalanges and there was the usual curving and dryness of the nails In the x ray there appeared the usual changes of advanced osteoarthritic destruction and deformation (Fig 64)

REFERENCES

1 Bickel W H Ghormley R K Coventry M D and Mussey R D *Proc Staff Meet Mayo Clin* Nov 1941
2 Brindley H *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *19 D* 105 1948
3 Browne H T *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *11* Y 49 1937
4 Cohan A Trommer T H and Goldman J *J A M A* *130* 765 1946
5 Dickson J A and Willien L J *J Bone & Joint Surg* *29* 687 1947
6 Ghormley R K *Proc Staff Meet Mayo Clin* Nov 1944
7 Haggart G E *J Bone & Joint Surg* *22* 717 1940
8 ——— *New England J Med* *236* 971 1947
9 Hall R F *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *18 D* 15 1946
10 Harmon P H *Surg Gynec & Obst* *76* 347 1943

with thickening and scar formation and sometimes even with ossification (Fig 65)

Gradually the inflammatory signs in the intervertebral articulations subside, and the condition becomes quiescent or entirely arrested This may occur before any ankylosis has taken place, or the process may go on to complete ankylosis Accordingly, one may make a clinical subdivision of the simple atrophic arthritis of the spine on one hand, and the ankylosing type on the other The atrophic arthritis is not confined to a single vertebra, but always occupies a section of the spinal column and sometimes the entire spine

B THE CLINICAL SYMPTOMS

1 Early subjective signs

The first sign referable to the spinal column is a certain occasional discomfort and pain at rest, which may disappear after some activity It is particularly noticed in the morning, and the patients are sensitive to weather changes Before the appearance of actual pain, paresthesia, tingling and numbness in certain portions of the body are not uncommon

2 Limitation of motion

Restriction of motion in different directions is noticed almost at once in the involved sections of the spinal column This limitation is at first entirely contractural, due to the protective spasm of the spinal muscles The greater the normal mobility of the spine, the more obvious is the restriction caused by this muscle spasm It will, therefore, be most noticeable in the cervical and lumbar portions In the dorsal spine, on the other hand, the paresthesias prevail The patient maintains a peculiar guarded attitude which is noticeable in his gait and in the ordinary movements of his body

3 Radiation

Radiating pain develops later It is characteristically circumferential in the dorsal spine, that is, it encircles the thorax, marking the course of the intercostal nerves In the lumbar spine, however, the radiation is directed toward the lower extremity and, according to the seat, may occupy the territories of the femoral, sciatic, ilioinguinal, or iliohypogastric nerves In the cervical spine the radiation occupies the territory of the occipital nerves and the brachial plexus Characteristically the radiation which is due to impingement of intraspinal nerves is not confined to the anterior primary division, but includes also the dorsal region of the body supplied by the posterior primary division of the spinal nerve This is to be expected, because the impingement occurs in or near the intervertebral foramen before the division of the spinal nerve

4 The deformity

Spinal deformity develops slowly and never reaches the degree seen in more destructive lesions The gradually developing obliteration of the joint may

Lecture V

ON SPONDYLARTHRITIS A NONSPECIFIC ARTHRITIS OF THE SPINAL COLUMN

THERE is no sound pathological basis for separating the arthritis of the intervertebral articulations from arthritis in general From the clinical point of view, on the other hand, there are several reasons why spondylarthritis occupies a place of its own First of all, the strict division into the hypertrophic and atrophic types is not always feasible, because a mixture of these two types occurs only too often In older age groups we observe frequently that the hypertrophic arthritis as a feature of senescence (Oppenheimer[1]) becomes superimposed upon an earlier rheumatoid type

Then also the proximity of the spinal nerves and their frequent involvement in the arthritic process is responsible for certain patterns of radiation which require careful analysis from the diagnostic viewpoint They must be differentiated from a multitude of other pain producing conditions not connected with the spinal column Failure to do so may lead to erroneous diagnoses, and worse yet, to faulty indications and sometimes even to unwarranted operations



Fig 65 Atrophic arthritis of the spine calcification of ligaments

I THE ATROPHIC ARTHRITIS OF THE SPINE

A THE PATHOLOGY

The changes which occur in the intervertebral articulations are quite identical with those seen in the larger joints They start in the synovial membrane of the intervertebral articulations as well as in the costovertebral joints (Klinge) There is the fibrinoid degeneration of the synovial membrane, and the production of an exudate and of a pannus, just as in the other joints In the end, an obliteration of the joint takes place by means of fibrous adhesions and often by complete bony ankylosis In addition to this, it is peculiar of spinal arthritis that the numerous periarticular ligaments likewise show cellular infiltration in the acute stages and finally end up

rigidity of the spine which starts in the dorsal region and extends downward, producing a characteristic dorsum rotundum Strumpell[16] and Pierre Marie[10] described another type of spondylitis also characterized by progressive stiffening which, however, begins in the lower portion of the spine and often includes the hip joint (spondylose rhizomelique)

In all types, the changes in the joints of the spine parallel those in the large articulations The same type of pannus formation occurs, which finally leads to destruction of the cartilage and lacunar erosion There is the same activity of the bone marrow which becomes fibrosed, invades the cartilage itself, and replaces it entirely



Fig 67 Ankylosing type of spondylarthritis Note bamboo like formation due to calcification of longitudinal ligaments

Attention should be directed especially to the appearance of arthritic changes in the sacro iliac joint It is one of the early signs of the atrophic spondylitis in general, and more particularly of the Strumpell Marie type In the so called Bechterew type, in which the lesion is confined to the thoracic and cervical vertebrae, we do not find the early sacro iliac changes In the Strumpell-Marie type these changes always precede any other x ray signs of spondylitis (Hare[5]), and in the early stages they can be recognized by the increased density of the bone adjacent to the articulation[4] (Fig 68) These are then followed by a total loss of structural detail, and finally the entire joint outline disappears At first the sclerosis of the sacro iliac joint is limited to the iliac bone, the sacrum becomes involved later



Fig 68 Atrophic pondylarthritis Note obliteration of sacro iliac joint Case A C

In contrast, in the hypertrophic arthritis the sacro iliac joint does not become involved in the process of ankylosis, nor do the other articulations of the spine If the sacro iliac joint appears effaced in the x ray picture, it is due to the oblique position of the joint at the sacro iliac junction (Borak[4]), and not to a true fusion Also, in the hypertrophic type the sacro iliac pain is absent, and the patient does not walk like one with sacro iliac disease, as is the case in the atrophic type (Scott[13])

produce a considerable degree of dorsum rotundum or a flattening of the lumbar or cervical spine But there is never the acute angulation which one sees in tuberculosis or, sometimes, in osteomyelitis

C THE X-RAY SIGNS OF ATROPHIC ARTHRITIS OF THE SPINE

1 In atrophic spondylarthritis

In the atrophic arthritis of the spine the discs are preserved much longer than in the hypertrophic type Nevertheless, with the progressing ankylosis the disc loses its function and later undergoes gradual degeneration The entire spinal column undergoes progressive atrophy, and the articulations become blurred and can no longer be definitely made out The contours are wavy and indistinct

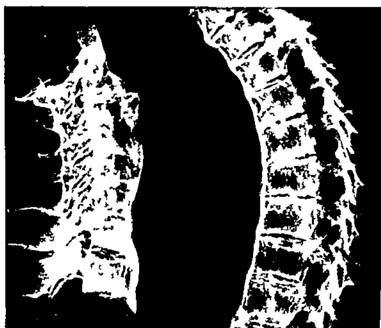


Fig 66 Spondylitis ankylopoietica or rhizomelique Note calcification of longitudinal ligaments

2 In ankylosing spondylarthritis or spondylitis ankylopoietica

This is merely a variant of the atrophic type Its characteristic feature is that it leads to early and complete ankylosis of the intervertebral joints The most striking feature, however, is the ossification of the ligamentous apparatus, involving both the long ligaments of the spinal column and the short ones (Fig 66)

In the x ray picture the spine assumes a characteristic bamboo appearance and the discs are thinned out and irregular The intervertebral articulations appear obliterated as do also the costovertebral joints (Fig 67) In the lateral view, one notices the ossification of the anterior longitudinal ligament There are two clinical subdivisions, the Bechterew and the Strumpell Marie types In 1893 Bechterew described a type of spondylitis characterized by progressive

are not the primary lesions In osteoarthritis of the spine the disease starts in the intervertebral disc, in contrast to the atrophic type of spondyl arthritis where the changes start in the intervertebral articulations

The first change in the disc is the fibrillation and the liquefaction of the hyaline cartilage with formation of fissures and cavities Large clefts appear This is then followed by calcification and granular decay of the fibers, with disappearance of the cells In keeping with the nature of hypertrophic arthritis, we see a revascularization of the cartilage layers from the adjacent cancellous bone of the vertebral bodies These secondary proliferative changes follow degeneration of the cartilage of the disc and result in the formation of exostoses, precisely as in hypertrophic arthritis of peripheral joints

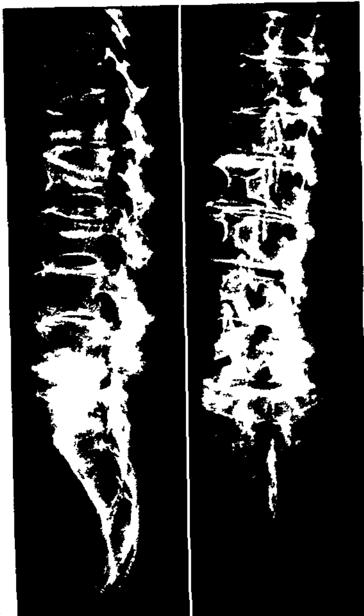


FIG 70 Osteoarthriti of the spine Bone bridges and spurs and degeneration of the disc

As a result of the degeneration, the disc loses all its physical properties It becomes an inelastic body and finally disappears entirely between the margins of the exostoses, which now form heavy ledges bridging the intervertebral space (Fig 69) All this conforms with the general picture of arthritis deformans It is peculiar, however, that the bone proliferations do not confine themselves strictly to the margins and circumference of the body, but they may also penetrate into the disc itself

The reason why these exostoses form mainly at the periphery are the tension stresses of the ligamentous apparatus, they develop in proportion to these stresses and fail to develop if no static stresses warrant their appearance The ossifications can be considered as physiological reactions to these stresses, necessitated by the degeneration and later the disappearance of the disc They are seen mostly at the anterior margins of the vertebral bodies, while the pos

As the spondylitis advances, the process of decalcification so characteristic of the atrophic type sets in the spine and later includes the sacro iliac joint which, as a result, loses its earlier sclerotic appearance

D THE DIFFERENTIAL DIAGNOSIS

1 Osteomyelitis

Osteomyelitis due to pyogenic bacteria or to the typhoid or paratyphoid bacillus shows early destruction of the intervertebral disc, in contrast to the atrophic spondylitis There are also structural changes in the body of the vertebra which are not observed in spondylarthritis In acute cases differentiation between arthritis and infectious osteomyelitis is not difficult, in subacute and chronic cases, however, the distinction is not always easy

2 Vertebral epiphysitis

The vertebral epiphysitis or the kyphosis adolescentium, the so called Scheuermann's disease, is a pure growth disturbance of the epiphyseal cartilage The narrowing of the disc which we see in this disease is not due to any destructive inflammatory process Nevertheless, secondary arthritic changes in the spine may appear after years in cases of Scheuermann's disease, but they are of the hypertrophic type

II THE HYPERTROPHIC TYPE OF SPONDYLARTHRITIS OR THE SPONDYLARTHRITIS DEFORMANS

A THE PATHOLOGY

So far as the intervertebral joints are concerned, the changes are essentially the same as in degenerative arthritis of the large joints But the joint changes



Fig 69 Hypertrophic spondylarthritis Note degeneration of disc and bone bridge uniting vertebral bodies

radiated to the anterior abdominal area one year prior to admission It was mistaken for gall bladder disease, and the gall bladder was removed The pain from the lower dorsal spine usually radiates around the body and is referred to the upper abdomen, simulating appendicitis or gall bladder disease The pain originating from the upper lumbar spine is usually distributed along the iliohypogastric, the ilio inguinal, and especially the external cutaneous nerves It is not unusual for this pain to appear bilaterally If the lower lumbar spine is involved at the lumbosacral junction, there is frequently sciatic radiation which may simulate intraspinal pressure produced by a herniated disc or by an intraspinal tumor The pain, however, is often bilateral, shows poor anatomical distribution, and rarely extends below the knees The anatomical reason for the occurrence of this radiating pain is undoubtedly the changes which occur in the intervertebral foramina The formation of small exostoses and the inflamma tory swelling of the synovial membrane produce a narrowing of the interverte-bral foramina and cause pressure upon the spinal nerves

III THE TREATMENT OF SPONDYLARTHRITIS (ATROPHIC AND HYPERTROPHIC)

Inasmuch as the treatment of the two types of arthritis does not vary es-sentially, there is no reason for a separate discussion of each type It seems to us more to the point to differentiate the treatment according to the phase of the disease, distinguishing particularly between the earlier phases in which pain is predominant and the later phases which are characterized by rigidity and deformity

A CONSERVATIVE TREATMENT

The conservative treatment of spondylarthritis consists of two components The basic treatment, which includes such general measures as sedation, gold therapy, hygienic treatment, etc , already discussed under general treatment for arthritis, and the local conservative treatment, that consists mainly in rest and recumbency, x ray therapy, local physiotherapy and im mobilization



Fig 71 Atrophic spondylarthritis (Strumpell Marie) Case W H (male) #40 9795 29 years July 1940 Low back pain and pain in both hips for many years increasing stiffness Acute recrudescence with marked muscle spasm requiring bed rest Treatment by traction and physiotherapy for 10 days patient able to leave bed and walk with lumbosacral support

terior borders are unusually free The posterior longitudinal ligament is thin and maintains its connection with the disc rather than with the vertebrae Occasionally we see small nodes of ossification in the ligamentum flavum The bone itself shows the same changes we see in the larger joints, namely cavities and subchondral cysts filled with blood, fibrin, or detritus of bone (Trummer cysts)

This illustrates from the pathological standpoint the contrast between the spondylarthritis deformans and the ankylopoietic arthritis The latter belongs to the atrophic type and starts in the articulation, while the hypertrophic arthritis starts in the disc and the articulations follow In the ankylotic type the ossification of the anterior longitudinal ligament which forms the so called bamboo spine is characteristic, but this should not be confused with the formation of true exostoses seen in hypertrophic arthritis of the spine (Fig 70)

B THE CLINICAL SYMPTOMS

1 Subjective symptoms

The earliest symptoms of the osteoarthritic spine are pain, stiffness, tenderness along the spinous processes paresthesia, radiating pain, and restriction of motion We find that the pain is less severe and more readily controlled in the hypertrophic type of arthritis It is aggravated by strain and stress on exertion and is at least for a considerable length of time, relieved by rest This type of arthritis involves people of middle age and older, whereas in the atrophic type, the younger age group is prevalent The disease develops slowly and insidiously, and the patients pay little attention to it sometimes for a number of years until the symptoms become exaggerated

2 Radiation in hypertrophic spondylarthritis

Several relevant points should be emphasized in relation to this most disturbing of all symptoms 1 There is no relation between the amount of changes seen in the x ray picture and the extent and intensity of radiation It usually takes in a number of sensory dermatomes and is seldom confined to a single one 2 On the other hand it never occurs in the strict territorial limitations which we see in the herniated disc 3 Furthermore, the radiation in the arthritic spine is accompanied by other sensory disturbances such as paresthesia, numbness, or burning pain It is therefore not of the purely protopathic neuralgic type which we see in other conditions Anesthesia is quite unusual, but hyperesthesia is not at all infrequent 4 The radiating pain usually precedes complete rigidity of the spine and often subsides later when the spine has become entirely immobile 5 The differential point is that the pain is usually relieved by positioning or lying down This is in contrast to the pain caused by a disc or a spinal tumor, where such relief cannot be obtained by positioning or rest alone

Radiation from the lower dorsal and upper lumbar spine often gives rise to erroneous diagnosis of an intra abdominal condition In one case the pain

to x ray are due to the liberation of sulfur within the body in such a form that it can replenish any local sulfur deficiency We have already remarked, however, that sulfur deficiency is not generally accepted as a factor in arthritis According to Hemphill and Reeves,[6] the value of x ray therapy consists in facilitating orthopedic correction and prevention of the deformity by reducing pain, stiffness, and paravertebral muscle spasm In their experience, pain limited to the sacro iliac joint was almost uniformly controlled by x ray In moderately advanced cases they state that 91 per cent were improved, whereas in farther advanced cases of ankylosed sacro iliac joints control of pain was obtained in 82 per cent An increase of motion was noted in 81 per cent in the early group, and in 69 per cent in moderately advanced cases, whereas far advanced cases showed improvement in motion in only 5 per cent Our experience with x ray treatment in the Strumpell Marie type of arthritis have been somewhat less favorable Of eight cases treated, one was an early case seen within one year after the beginning of the complaint, two were seen within two years, one after three, and three after four years Of these eight cases, seven or 87 5 per cent had immediate relief of pain, and three or 43 per cent had increased mobility also One patient, followed as long as one year, had no recurrences However, no result was obtained in cases of very long standing

A combination of x ray and orthopedic treatment of the Strumpell Marie arthritis is advocated by Baker [1] The x ray treatment is given early for the alleviation of pain and muscle spasm This allows more rapid and more effective correction of the deformity which is accomplished by special exercises designed to develop the muscles of the shoulder girdle, to expand the thoracic cage, as well as to strengthen the abdominal wall The posture of the patient is improved by proper exercises and with the assistance of a shoulder brace In patients treated during the early stages, encouraging results are obtained with this combined method which reduces pain and stiffness, increases spinal motion and chest expansion, and releases the paravertebral muscle spasm

B THE OPERATIVE TREATMENT OF SPONDYLARTHRITIS

We have learned long since that forcible correction of the deformity is a dangerous and disastrous procedure, because it may lead to immediate destruction of the spinal cord It may be surprising to learn, therefore, that recently operative measures have been brought forth to improve the deformities These operations are based largely upon the concept that it is not the bodies themselves, but rather the intervertebral articulations which prevent correction of the deformity Smith Peterson, Larson and Aufranc[15] proposed an appropriate operative procedure for the correction of kyphosis in the ankylosing type of spondylarthritis They expose the posterior arches by a midline incision, reflecting the intraspinous and supraspinous ligaments, and splitting *and excising* the spinous processes at their bases Then, by subperiosteal dissection, both the superior and inferior articular facets of the intervertebral joints are exposed,

1 Recumbency and traction

Recumbency and traction is indicated in the most acute stages or in periods of acute recrudescence It should be applied for short periods only, rarely over more than one or two weeks, because the general health of the patient demands ambulation at the earliest possible moment (Fig 71)

2 The immobilizing ambulatory treatment

This is best carried out by means of braces rather than casts, so that the muscular spasm can be relieved by application of heat and massage When this spasm has been controlled, it is possible to obtain some correction by active exercises, but the back brace must still be used to rest the muscles during the intervals

3 The physiotherapy treatment

The physiotherapy treatment consisting in heat and massage and graded exercises should be applied as early as possible, whether the patient is recumbent or ambulatory

4 Correction of deformity by casts

While the disease is active, the flexion deformity of the spine increases, sometimes at a rapid rate The question then arises whether the spine is still extensible enough to undertake some corrective treatment The extensibility of the spine can be tested by short periods of head and pelvic traction in recumbency before one undertakes a more active regime of correction by means of casts A method based upon Telson's principle for the correction of the juvenile kyphosis is to place the patient in a plaster cast which reaches up to the apex of the curve in the back, and then to force the shoulders back into hyperextension *by means of straps fixed to a posterior iron cross piece*

In most cases of long standing, however, one will find that the condition is stationary and does not yield to mechanical force, the problem is then to prevent the progress of the deformity by immobilization For this purpose a properly constructed brace is sufficient in most cases There are, however, a few with a tendency to increasing deformity and with marked radiation symptoms, which will require a plaster cast for short periods, to be duly alternated with braces and physiotherapy

5 The x ray treatment

Initial and early stages are favorably influenced by x ray treatment if given in small doses, gradually covering the entire extent of the spine, the so called wide field application This treatment is more effective in the atrophic than in the hypertrophic type In the earlier stages the mobility may actually be increased In later stages one cannot expect an actual loosening of the spine from the treatment, all one can look forward to is the relief of pain Blair[3] *believes that the beneficial results obtained from wide exposure of the body*

B THE X-RAY FEATURES

In order to show the uncovertebral osteophytes and the marginal proliferations of the lower and upper vertebrae as they project into the intervertebral foramina, oblique views of the cervical spine should be taken Occasionally one can see an exostosis in these foramina The atlanto occipital and the atlanto odontoid articulations can be seen best in the anteroposterior film taken through the mouth (Fig 72)

In the atrophic type one notices irregularities in the articular outline and blurring of the joint contours, while the atrophic bodies of the vertebrae show increased transparency together with calcification of the ligaments In the hypertrophic type one also sees irregularities in the contours of the articulation, they appear wavy and show the characteristic subchondral sclerosis In addition, there are spurs and exostoses around the articular facets



Fig 72 Cervical spine atlanto odontoid articulation

The salient points in the diagnosis are For the atrophic type, the irregularity of the articulation and the atrophy of the vertebral bodies with increased transparency, and the calcification of ligaments For the hypertrophic type, the irregularity of the outlines of the articulations which, however, are not obliterated, the subchondral sclerosis, the spurs and exostoses around the articular facets, and the marginal exostoses and bone bridges which span the anterior borders of the vertebral bodies

C THE CLINICAL PATHOLOGY

The symptoms of cervical arthritis may be classified into two groups

1 Spastic rigidity and restriction of motion

Spastic rigidity is noted in the deep and short muscles of the neck, the rotators and extensors of the head, and in the more superficially located splenius and trapezius muscles The spastic muscles appear hard and indurated on palpation and are very tender The stethoscope reveals harsh sounds, even cracks, coming from the rubbing of the fibrosed soft tissues Under traction or under anesthesia these spastic muscles relax The spasticity is the cause of the restriction of motion The rigidity is not uniform, the motion between occiput, atlas, and odontoid usually remains free The spastic rigidity of earlier stages is amenable to mechanical treatment In the later stages of osteoarthritis the fixation of the head is due to skeletal changes, and the therapeutic problem is quite different

after the ligamentum flavum has been detached The articular processes of the vertebra involved, both the upper and the lower, are osteotomized obliquely In addition, the vertebra itself is gouged out through the pedicles for a distance of two or three spinal segments Then the head and foot parts of the operative table are raised so as to angulate the spine and to encompass the correction of the deformity The operation is completed by fusing the spine The rationale of this procedure is that the obstacle to the correction of the flexion deformity lies principally in the ankylosed facets covered with bony overgrowth The authors presented 6 cases, 5 in the lumbar and 1 in the thoracic region, all with satisfactory results The operation is most difficult and requires a master surgeon Judgment on this procedure must still be reserved

In a still more complicated procedure which has been suggested lately, *LaChapelle*[7] *goes so far as to osteotomize the lumbar spine for correction of* the kyphosis The operation is performed in two stages One is removal of the lamina and the posterior articulation, usually those of the 2nd and 3rd lumbar vertebrae, the second stage, performed through an anterior approach, consists in excision of the 2nd lumbar intervertebral disc The operation is extremely difficult and dangerous, and it is very doubtful that it will ever gain general acceptance

IV THE CERVICAL SPONDYLARTHRITIS

This localization of the spondylarthritis offers so many difficulties both in diagnostic and therapeutic respects, that it may well be discussed as a separate entity

A THE ANATOMY OF THE CERVICAL SPINE

Under normal conditions the superior surface of the cervical vertebra has two well marked lips which cause the body to appear concave from side to side, as seen in the anteroposterior picture The inferior surface, on the other hand, is bevelled so that it appears convex in the same view

If, however, we view the joint from the side, we will find that the upper surface of the vertebra is convex, while the lower is concave In other words, the upper surface is concave in the frontal and convex in the sagittal plane, while the lower surface is convex in the frontal and concave in the sagittal plane This means that each adjoining vertebral surface has a double curve in opposite directions

Furthermore, one can see that the intervertebral foramina from the 3rd to the 7th cervical vertebrae are bounded by the lateral lips of the superior surface of the lower vertebrae and the bevelled lateral margin of the inferior surface of the upper vertebrae According to Luschka,[8] there is often a very *small articular body between these uncinate processes and the laterally bevelled* border of the adjacent vertebra which forms a hemiarthrosis with the vertebra (Luschka[8]) Lyon[9] believes that these formations cause the neuralgic symptoms, and he calls the condition spondylosis deformans of the intervertebral hemiarthrosis

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middle cord, that is, into the territory of the median nerve, and usually involves the index and middle fingers but rarely the thumb Again, if the 5th or 6th cervical roots are involved then the radiation will go down the shoulder to the upper arm, particularly to the deltoid region

D THE DIFFERENTIAL DIAGNOSIS

There are so many different conditions capable of producing pain radiation in the same territories as the arthritis of the cervical spine that it would lead us too far afield to consider every single diagnostic possibility The most frequent conditions requiring differentiation are the following

1 *Cervical disc*

The most common differential problem is the pain produced by the rupture of an intervertebral disc of the cervical spine or by an intraspinal tumor These patients complain of pain in the occiput, *neck, shoulder, arm, and* fingers

There are two clinical groups One is the acute variety with sudden onset which follows definite trauma It includes the so called explosive type of rupture of the intervertebral disc which is found in patients injured in diving or automobile accidents Differentiation is not difficult where there is a clear-cut history and where the condition is unilateral The diagnostic difficulty lies more in the so called central type of herniated disc where the spinal nerves may be involved on both sides The patient complains of stiffness of the neck, and there are sensory and reflex changes, weakness of muscles, and not in frequently vasomotor disturbances There is the same loss or reversal of the *normal* cervical lordosis which we see in osteoarthritis, but in contrast to the latter, relief is not obtained by rest or traction

The second group is the chronic type of cervical disc which follows mild or repeated trauma (Semmes[14]) The patients complain of cricks in the neck, with pain in the occiput, shoulders, arms, fingers, and even in the precordial regions, and also numbness and paresthesias of the fingers, as is the case in cervical arthritis In fact, these cases are often diagnosed as neuritis, tendinitis, myositis, or scalenus anticus syndrome There are, however, some differential points In the case of a disc, downward pressure on the head is likely to increase the pain, it is not relieved by traction as in arthritis, and is not particularly influenced by positioning the head The *Naffziger sign* is positive

2 Syringomyelia

Another condition which sometimes requires differentiation *is syringomyelia* Symptoms of this condition are radiating pain and *paresthesia*, but the difference is that the extent of the lesions is wider than in the cervical *arthritis*, and they do not follow any anatomical distribution There is usually the charac-

2 The pain

a The Local Pain

It depends upon the particular structure involved whether the pain appears strictly localized or diffuse If it is caused by the tenderness of the spastic muscles, it is not sharply localized and there are no distinct pressure points A small circumscribed pain area indicates its origin to be in the ligamentous and capsular apparatus of the intervertebral articulations In this case discrete pressure points are present Pressure upon the transverse processes of the cervical vertebrae is usually painful, especially at the 6th cervical transverse process, the so called tubercle of Chassaignac The tenderness of compressed roots of the brachial plexus is noted at the point where the spinal nerves emerge in front of the transverse processes in the lateral triangle of the neck

b The Radiating Pain

The most common patterns of radiation in cervical arthritis are the following

1) Occipital radiation It covers the territory supplied by the greater occipital nerve The nerve leaves the 3rd intervertebral foramen in the triangle formed by the superior and inferior obliques and the rectus capitis posterior major Pressure in this region will cause radiation of pain over the occiput, the partietal and even the frontal bone The nerve comes from the posterior division of the 3rd cervical nerve Its involvement indicates arthritis of the upper cervical spine

2) Shoulder girdle radiation The radiation extends along the trapezius, deltoid and pectoral regions It is the territory supplied by the anterior and posterior divisions of the 4th to the 7th cervical nerves The territory of the anterior primary division, reaches farther distally over the region of the deltoid and pectoralis major The skin over the trapezius in the back is supplied by sensory nerves which come from the posterior primary division of the 4th cervical segment As is to be expected, the pain manifestation will involve both anterior and posterior primary divisions, because the seat of irritation is in the intervertebral foramina, that is, before the nerve bifurcates into its primary divisions (Nachlas[11])

3) Radiation into the brachial plexus When the arthritis involves the lower half of the cervical spine and the 1st and 2nd dorsal vertebrae it produces radiation in the arm and forearm following the distribution of the cervicodorsal plexus The upper part of this plexus consisting of the 5th and 6th cervical roots forms principally the posterior cord The middle portion, the 6th and 7th cervical is the territory of the median nerve, the lower cord, the 8th cervical and 1st dorsal, is the territory of the ulnar nerve Consequently, involvement of the lower cervical spine, will produce radiating symptoms in the territory of the ulnar nerve and the cutaneous antibrachii medialis On the other hand, if the 6th or 7th cervical is involved, then the radiation extends into the

and rate of progress of the arthritis which has existed before the trauma, and then upon the prompt appearance of the signs of aggravation after the trauma The symptoms must be appropriately localized, and the x ray changes must show the presence of a degenerative arthritis To a certain extent, we can rely upon the continuity of symptoms together with the assurance that the complaints actually started with the accident, and then carried on in a continuous chain up to the time of examination Such a combination would indicate that the trauma was actually related to the arthritis

6 The scalenus syndrome and the cervical rib

The pattern of radiation in the cervical rib case may simulate osteoarthritis In the latter the radiation is more frequently bilateral, in contrast to the cervical rib Cervical arthritis in the painful stage is almost uniformly benefited by head traction in the proper direction, in the cervical rib, as well as in the scalenus syndrome, elevation of the arm is more likely to bring relief The presence of very definite pressure points at the brachial plexus as it is pressed against the cervical rib and, in the last analysis, the x ray picture will decide the diagnosis

E THE TREATMENT OF CERVICAL ARTHRITIS

1 Elimination of the cause

Only in a small number of cases is it possible to establish a focus of infection as the cause of the arthritis Such a focus is usually situated in the upper air passages, the teeth or tonsils, or the paranasal sinuses We have observed a number of cases in which the extraction of a tooth or the drainage of a paranasal sinus has brought considerable relief Although these cases are rare, they impose upon the surgeon the obligation to make a careful search for focal infections in the upper air passages

2 The supportive treatment

The supportive treatment is a necessary adjuvant in all kinds of therapy, whether the latter involves the elimination of a focus or any other means It consists in rest, good elimination, proper diet, and enough sedation to assure the patient an adequate amount of sleep The general resistance of the patient must be raised under all circumstances He should be kept on a liberal high caloric, well balanced diet, with emphasis on proteins rather than on carbohydrates The sedatives are preferably given in form of barbiturates

3 The symptomatic treatment

a Conservative Treatment for the Relief of Pain

As pain is closely related to motion, and as it responds to rest and immobilization, appropriate measures should be taken The arthritic pain responds almost invariably to traction Such traction should be applied during the examination

teristic disturbance of thermic and pain sensations The following is a case in point

Case B B (Male) #62 019
Age 52 years Adm November 12, 1947

This patient complained of occasional pain in the neck radiating to the left shoulder and down the arm to the fingers, for eight years For the last two months he complained of burning pain in the left shoulder which spread to the left hand and extended down the chest On examination we found a slight narrowing of the left palpebral fissure with atrophy and weakness of the left upper extremity There was absence of the left triceps jerk A zone of diminished sensation was noted over the outer aspect of the left arm, shoulder and forearm The x ray showed a narrowed space between the 5th and 6th cervical vertebrae and some arthritic lipping, which did not explain the clinical picture This case was finally diagnosed as syringomyelia He was treated with deep x ray to which he responded well The pain in the neck disappeared although the sensory changes of paresthesia and hypesthesia remained

3 **Fibrositis and myositis**

It is rather difficult to establish a differential diagnosis because these conditions are simply additions to the clinical picture of arthritis, and myositis of the neck muscles may be considered as part of the arthritic syndrome The myositis manifests itself by susceptibility to weather changes, to moisture, and to exposure to cold and damp weather The muscles are in condition of spasm, show signs of early fatigue and, objectively, some inflammatory hyperplasia of the connective tissue Even serofibrinous exudate with proliferation of fibroblasts and blood vessels has been reported Clinically, local muscle indurations and sometimes subcutaneous nodes are observed

4 **Scapular grating**

It is often caused by a localized interstitial myositis of the shoulder girdle, although there are other factors which may produce the grating, such as exostoses, bursae, of even congenital anomalies of the thorax There is usually a very definite pressure point at the upper inner angle of the scapula

5 **Trauma and cervical arthritis**

It is entirely irrelevant to make a distinction between so called traumatic and hypertrophic arthritis because, pathologically, the two conditions are identical The question is, whether trauma plays any role in the production of arthritic symptoms or in their aggravation Any definite trauma affecting the cervical spine, such as a contusion of the articulation or compression of the intervertebral disc or even fractures of the articular processes, may easily lead to exacerbation of an already existing degenerative arthritis Since hypertrophic arthritic changes are encountered in normal life and are particularly frequent in certain age periods, the distinction becomes difficult If one is to decide how much the symptoms are due to the pre existing arthritis and how much to an accident, judgment must be based first of all upon the course

may be repeated several times a day Between treatments the patient is allowed to rest, after which stretching is resumed again As soon as the patient has been relieved by bed traction or intermittent traction, we apply a collar, usually of the type of the so called neck extension brace Exercises are instituted together with massage from the beginning These exercises consist in active rotation, flexion and extension movements, and are supplemented by careful passive, resistant exercises Massage should always be preceded by heat We prefer the infra red light because it is most easily applied and is also the most effective

b Operative Treatment for Pain

Is operative treatment ever indicated? In the course of time the spine becomes automatically stabilized by a true ankylosis or by the locking of the intervertebral joints There is, therefore, hardly ever the need of securing stability of the spine by operative measures We have never found it necessary to carry out spinal fusion for this purpose

c Treatment of the Deformity

The treatment during the acute and subacute stages should be directed toward the prevention of deformity Deformity cannot be changed once it has fully developed Fortunately, in cervical arthritis the deformity is never pronounced, except in those cases of the ankylosing or Strumpell Marie type which end with strong forward flexion of the head and which handicaps the patient in walking We have found on several occasions that a good deal of limbering up can be accomplished by x ray treatment, followed by traction and immobilization

REFERENCES

1 Baker L D *Arch Phys Med* 26 389 1945
2 Bechterew V M V *Neurol Zentralbl* 502 426 1893 *Deutsche Ztschr f Nerven heilk* 9 1897 and 15 1899
3 Blair H C *Surg Gynec & Obst* 74 663 1942
4 Borak J *Radiology* 47 128 1946
5 Hare H F *New England J Med* 223 702 1940
6 Hemphill J E and Reeves R J *Am J Roentgenol* 54 282 1945
7 LaChapelle E H *J Bone & Joint Surg* 28 851 1946
8 Luschka H v *Halbgelenke des Menschlichen Korpers* Berlin 1859 4
9 Lyon E *J Bone & Joint Surg* 28 851 1946
10 Marie Pierre *Rev de Med Paris* 1898 *Semaine Med* 1899
11 Nachlas I *South Med J* 35 665 1942
12 Oppenheimer A *Am J Roentgenol* 49 1 1943
13 Scott G *A Monograph on Adolescent Spondylitis or Ankylosing Spondylitis* London Oxford Univ Press 1942
14 Semmes R E *Am J Surg* 74 137 1948
15 Smith Peterson M N Larson C B and Aufranc O E *J Bone & Joint Surg* 27 1 1945
16 Strumpell A v *Deutsche Ztschr f Nervenheilk* 11 1897

to test its effect The patient will unfailingly express a feeling of relief if the traction is carried out in the proper direction This is due to the fact that the spastic, contracted muscles are being stretched and, also, that there is relief of the pressure upon the spinal nerves In bed, this traction can be carried out very easily by means of a head sling to which 6 to 9 pounds are attached The head end of the bed is raised so that the patient's own weight furnishes the countertraction Traction must be directed with the head in proper position, either slightly forward or slightly backward, according to what the patient

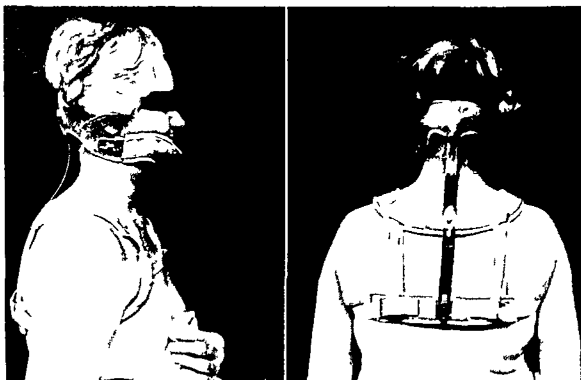


FIG 73 Neck extension brace

states is most comfortable Most patients will be relieved within a few days, sometimes within a few hours

When the tenderness has disappeared to the extent that the patient can move in bed without any pain and when the pain does not recur as soon as traction is discontinued, he is ready to leave the bed He is then placed in immobilizing apparatus A patient can easily be furnished with a so called neck extension brace such as we use in our clinic (Fig 73) The brace is worn continuously at first, as long as the patient is on his feet, later the collar may be taken off for various periods of time, according to the tolerance of the patient From the beginning, physiotherapy consisting of heat and massage is applied to the painful areas

The *plan for the systematic treatment*, therefore, formulates itself as follows In the acute stages it consists in traction in bed with application of heat In *subacute and chronic stages* one applies intermittent traction by a Sayre sling or suspension apparatus while the patient is seated in a chair This procedure

theory was finally corroborated by Ebstein,[3] who devoted considerable time to experimental studies and became a great exponent of the view that the necrotic changes in the affected tissues, are the primary cause, and that only following this necrotising process in the cartilage and other implied structures is uric acid deposited into the necrosed areas

We may therefore define gout as a disorder of the metabolism associated with the retention of uric acid and other purine bodies, and characterized clinically by attacks of acute arthritis and by the deposition of sodium biurate in and about the joint The purine bodies which accumulate are adenine, guanine, and xanthine, as well as uric acid, and they all come from nucleoproteins of food as well as of the tissues, being formed by means of a certain fermentative enzyme

The normal daily output of uric acid through the urine is 0 4 to 1 5 gms This amount, however, is greatly influenced by food, particularly food rich in purine bases For instance, after a meal containing sweet bread the amount of uric acid output may be doubled At the same time, an excess of uric acid is found in the blood, in fact, the content may rise as high as 10 mgm per cent and even more There has been some speculation as to the reason why the blood is overcharged with uric acid, whether it be that there is a normal production and a deficient excretion of uric acid, or that there is an overproduction and a normal excretion Observers now agree that the presence of uric acid in increased amounts does not indicate a deficient excretion in the urine or a diminished destruction of uric acid by imperfect oxidation, but rather, an overproduction of uric acid with normal excretion Up to 1859 it was supposed that this uric acid was formed in the kidney itself In that year Sir Alfred Garrod[4] demonstrated the presence of uric acid in the blood of gouty subjects, and this led to the conclusion that the uric acid was formed in certain organs and tissues of the body and was merely eliminated by the kidneys Garrod himself came later to the conclusion that uric acid is produced by direct action of the kidneys alone Others such as Schroder and Minkowski have demonstrated that the liver and spleen also produce it Ebstein[3] found uric acid in muscle and bone marrow of the affected extremity, he believed, though, that the kidneys take part in both the excretion and the production of it

Be this as it may, we must consider whether uric acid is a normal constituent of the blood Here again we find the statement of Garrod,[4] that in absolute health uric acid is present in the blood in only small quantities, while gouty blood is very rich in it We know now that there is a normal level in the blood of 1 7 mgm per 100 cc , and that this level is considerably increased in gout This excess of uric acid, however, is by no means characteristic of gout alone, because it is also found in other conditions such as leukemia and renal disease [7] Consequently, we may expect that the urinary output of uric acid may be either normal or low, and that the uric acid is retained in the system In fact, there are cases of gout in which the excretion of uric acid through the kidneys has ceased entirely in the intervals between attacks

Lecture VI

ON GOUT (ARTHRITIS URICA)

WE LEARN from the Father of Medicine on the subject of gout that "the podagra is the most violent of all joint diseases, it has the longest duration, and is the most difficult to treat"[6] The Greek physicians recognized the conditions and distinguished between the podagra of the feet, the chiragra of the hands, the gonagra of the knee, etc, but gout and rheumatism were to them one condition, and they used these terms indiscriminately

It was not until the 17th century that a differentiation was made between the two diseases The early classic on gout as a clinical entity sprang from the pen of Sydenham in 1683,[13] but more than 100 years elapsed before the systemic nature of the disease was recognized Scudamore, in 1817, described it as a constitutional disease, producing external local inflammation and based upon hereditary or constitutional factors even though acquired during life According to Luff,[9] it is a manifestation of a number of morbid tendencies, some inherited and some acquired, which result in a certain arthritic diathesis

I THE PATHOGENESIS OF GOUT

A THE ROLE OF URIC ACID

Gout is associated with an excess of uric acid in the blood The question is, how is this excess produced, how is it introduced into the blood, what is the source of its production, and what is its relationship to the gouty paroxysm and to other manifestations of the disease?

In former years the excessive uric acid found in the blood was considered the primary cause It was believed that the uric acid compound either acts mechanically while in crystalline state or works in solution as a poisonous irritant A proponent of the former view was Sir William Roberts,[12] who considered the disease a manifestation of mechanical injury produced by uric acid crystals The other view, namely, that the uric acid compound acting in solution is a poison, is equally untenable because there is no direct experimental evidence that uric acid is a toxic agent Although the fluids of the patient may be impregnated by sodium biurate to saturation on the eve of the outbreak of gout, yet no signs of poisoning can be noticed

The theory we now accept is that gout is a disease due to morbid changes in the tissues, and that these changes are the primary condition It is they which cause the necrosis and not the presence of dissolved urates Ord, in 1872, was the first to consider gout as being due to a special form of degeneration of some of the fibroid tissue which results in excessive formation of sodium biurate This is then discharged into the blood and subsequently is deposited into those parts which are least freely supplied with vascular and lymphatic structures This

degeneration, which reduces its elasticity and coherence and leads to folding and wrinkling

B THE SYNOVIAL CHANGES

The synovial membrane shows a marked reaction Here also white punctate foci of chalky deposits are seen, especially in thickened villi The synovia produces a pannus in form of a fine membrane extending over the cartilage The pannus finally destroys the cartilage and leads to a fibrous ankylosis

Other changes appear in the synovial villi which contain deposits of uric acid, as does the synovial membrane itself Nests of urate deposits can be seen



FIG 74 Urate crystals in superficial layers of cartilage (From Lange *Handb d Spe Pathol Anat u Histolog* Berlin O Lubarsch and F Henke 93 32? 1939)



FIG 75 Projecting urate nodules in synovial membrane (From Lange *Handb d Spe Pathol Anat u Histolog* Berlin O Lubarsch and F Henke 93 3?6 Fig 18 1939)

everywhere in these synovial proliferations They are surrounded by cellular connective tissue and sometimes contain giant cells Masses of urates, if lodged close to the surface, will cause projecting nobs or nodules (Fig 75)

Finally, the cartilage is entirely substituted by synovial panus, and it is this pannus which forms the adhesions with its own blood vessels which it receives from the synovial membrane The subchondral marrow spaces do not seem to take any part in it

C THE CAPSULAR CHANGES

The capsule and ligaments are likewise involved in pathological changes, although here the urate deposits are very much less marked There may be some in the fibrous capsule or in the periosteum, but if so, they appear only in small quantities On the other hand, the fibrous capsule takes on some inflammatory thickening and fibrosis

D THE BONY CHANGES

At the borderline between the bone and joint cartilage we notice tears and fissures brought about purely by mechanical stresses, as the cartilage loses its

B THE PREDISPOSING FACTORS

It is generally accepted that there are predisposing factors operative in gout For one thing, the hereditary tendency is most important As many as 75 per cent of all cases show evidence of familial occurrence (Luff[9]) It is peculiar that the females in gouty families frequently escape the disease, but they transmit the susceptibility to their children Whether this transmission is of the dominant or the recessive type is still a question Alcohol, especially in the form of concentrated liquors, and food also play an important role, especially rich food of high purine content such as heavy beef or bacon and glandular food such as sweet breads, liver or kidney It has also been observed that attacks of gout in patients so predisposed may be precipitated by injury Obese people when put on a rigid starvation diet may develop gout Tuohy[14] reports a patient 70 years old, weighing 210 pounds, who was put on an 800 calorie diet, and in eight days developed gouty arthritis

Gout may also be precipitated medicinally, particularly by diuretics There are many observations to show that an ordinary spontaneous gout attack is preceded by an increased excretion of sodium chloride and water with consequent disturbance of the electrolytic balance Of the diuretics, those of the mercurial type are especially prone to provoke a gout attack and should not be given to patients with a gouty history Price[11] reports five cases in which there was a history of frequent gout attacks, and all patients developed copious diuresis with reduction of the edema following mercurial therapy In all five cases there occurred acute attacks of gout a few days after the maximum diuresis was reached, which was in seven to nine days One patient died from congestive heart failure within six weeks indicating that salyrgan, a mercurial diuretic, can be dangerous to this type of case Persons who early in life suffer from uric acid gravel are prone to develop gout later, because deposition of the acid in the kidneys produces pathological conditions which interfere with its proper elimination (Luff[9]) Finally, it is well known that gout attacks may be precipitated by psychic factors such as powerful emotion, fits of anger, worry or anxiety

II THE PATHOLOGY

A THE CARTILAGINOUS CHANGES[8]

The most important and the earliest changes are seen in the joint cartilage They appear as white chalky deposits sprinkled along the surface and the interior of the cartilage mass The deposits are situated mostly in the superficial layers and seem to decrease toward the depths Since the cartilage shows a maximum of sodium ion its preference for the deposit of urate is understandable Under the microscope these urates appear as needles in the superficial layers, gradually penetrating into the depths (Fig 74), although the manner of deposition seems to have no particular relationship to the architectural structure of the cartilage itself [10] The gouty cartilage undergoes colloid or mucoid

degeneration, which reduces its elasticity and coherence and leads to folding and wrinkling

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At the borderline between the bone and joint cartilage we notice tears and fissures brought about purely by mechanical stresses, as the cartilage loses its

elasticity The urate deposits, in the meantime, have penetrated from the superficial to the deeper layers so that they finally reach the zone of calcified cartilage

Nothing can be seen in the underlying bone of any activity of the marrow tongues, and *nothing of any penetration of these tongues into the cartilage,* as we see it in arthritis deformans The essential change in the bone marrow is that it undergoes a fibrous degeneration Within this fibrous marrow certain absorptive and reactive changes take place For one thing, we see tophi and uric acid appear in the deeper layers of the spongiosa Sometimes they reach beyond the neighboring zones of fibrous marrow and may penetrate even into the fatty *marrow beyond Side by side with this there are resorptive changes in the form of lacunar absorption Numerous giant cells are seen, presumably* attempting to remove the nodes of urates, and under favorable conditions the nodes may be absorbed, and may entirely disappear

Reactive changes of the periosteum appear in the form of exostoses, but there is no real gouty periostitis The exostoses usually show some static orientation *in their structure*

E THE CHANGES IN THE TENDONS

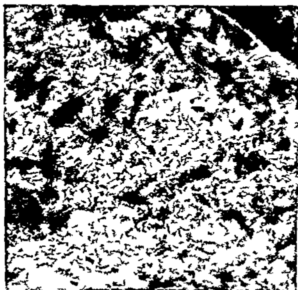


Fig 6 Tophus large masses of sodium biurate crystal

The changes in the tendons are less pronounced We note here also uric acid crystals deposited between the tendon fibrils, while the cells of the tendon fibers still show good staining qualities Therefore, we must assume that there is no preceding necrosis in the tendons The result of these deposits, however, provokes a reaction in the tendon itself which leads to edema and loosening of the coherent tendon fibers, as well as to reactive proliferation This changes considerably the physical quality of the tendon, particularly at the tendon insertion

F THE TOPHI

They are accumulations of urates under the skin, which they may erode, causing ulcers and sinuses These tophi consist of 60 per cent uric acid in the form of crystallized needles, usually easily soluble (Fig 76)

III THE CLINICAL PATHOLOGY

Comparatively little has been added to the *classic description of gout* by Sydenham (1683) It is a constitutional disease which produces inflammatory

changes of a specific kind, and which occurs most frequently between the ages of 25 and 40 It is now generally recognized that gout is more frequent than was usually believed It depends, of course, upon what the diagnosis of gout is based and how accurate it is One will find the disease not so infrequent if one considers that it is not only a joint disease with a specific characteristic textbook appearance, but also with cutaneous, renal, and parenchymatous ramifications Beitzke,[4] for instance, found among 100 pathological specimens of cadavers, eight afflicted with gout Heine,[5] examining 1000 anatomic specimens, found gout in 1 1 per cent It is eminently a disease of the male sex, the ratio of males to females being 91 9

The disease most frequently attacks the small joints of the hand and foot, hence the names, chiragra and podagra Less frequently the larger joints, such as knees, elbows, and shoulders are affected The disease occurs in acute and in chronic form

A THE ACUTE GOUT

In acute gout, the attacks occur as a rule in the early morning hours, the patient being aroused by severe pain, usually in the metatarsophalangeal joint of the big toe It is the typical location though by no means the only one, and it gives gout its characteristic stigma of big toe disease This joint swells rapidly, becomes hard, tense and shiny, and extremely painful and sensitive, the superficial veins become dilated A characteristic edema appears around the joint which distinguishes the condition from the rheumatoid arthritis The fever may reach 102 to 103 degrees, and during the acute phase there is some leukocytosis After some hours, the pain, which is sometimes accompanied by profuse perspiration, partially abates The attack may recur one or two nights after, with the same excruciating pain and local redness The acute stage usually lasts from one to two weeks, although sometimes the attack is prolonged in older patients

Gradually the acute symptoms of redness and pain subside, although the edema may linger on for some time As the swelling goes down, the skin desquamates, and finally a complete remission follows The first attack is more likely to occur during the winter or the beginning of spring It seems that one sees these typical classical attacks today less frequently than in the past, due possibly to greater restriction in eating and drinking, or to the better cultivation of healthy outdoor exercise With an increase in the frequency of attacks, the articular symptoms persist, and gradually many joints may become affected

Ankle Gout

Case R M L (Male) #34 044
Age 39 years Adm June 30, 1936

Pain in the right ankle with redness and swelling, appearing suddenly at two o clock in the morning and disappearing after a short time After one year of freedom, he had another episode, appearing at five o'clock in the morning On

admission the ankle was swollen and tender but not red The uric acid examination showed 6 5 mgm per cent

Big Toe Gout

Case L M P (Male) #39 561
Age 43 years Adm September 1, 1938

Eight days before admission the patient had severe pain in the left great toe The toe was red and swollen The examination showed swelling and tenderness over the metatarsophalangeal joint of the great toe The x ray picture was characteristic, showing a punched out area at the head of the metatarsal In this case the uric acid was as high as 7 2 mgm per cent



Fig 77 Gout urate deposits in tendon sheaths Case C C

B THE CHRONIC GOUT

Chronic gout develops insidiously from recurring attacks as they become more frequent and more prolonged, lasting for weeks and even months Deposits of urates appear in the cartilage and later in the ligaments and capsule, in the course of years the whole joint becomes swollen and deformed First, the hands and feet are involved, in severe cases there are extensive deposits about the elbows and knees Similar deposits are also found in the tendons and bursae (Fig 77), especially gouty bursitis is quite common

Urate Deposits in the Tendo Achillis

Case A L L (Male) #49 792
Age 53 years Adm December 2, 1942

Three years ago he experienced sudden pain in the big toe, diagnosed as gouty arthritis This finally subsided, and he was without pain until three months before admission Then he complained of having pain in the heel without any previous injury This pain subsided to return in two or three weeks At first the pain was nocturnal only The patient also stated that he had kidney stones three years previously The x ray showed small areas of absorption in the left os calcis,, and on both sides there was a deposit at the tendo Achillis insertion The blood uric acid was 3 8 mgm per cent

Tophi appear in various localities, most frequently in the cartilages of the ear and in the ends of the fingers The tophi consist in sodium biurate deposited under the skin They may comunicate with the joints and cause them to become considerably enlarged, although most of the distention is due to the thickening of the synovial membrane Other depositories of the urates are the kidneys, the presence of urates indicates that kidney function is inadequate

In these cases, a most careful examination of the urine should be made It is usually increased in amount, has low specific gravity, and contains a slight amount of albumin with a few hyaline casts In one of our cases (A P) the blood uric acid was 7 7 to 9 mgm per cent, and there was definite kidney damage with albumin and granular casts in the urine As the kidney condition becomes aggravated, other complications may follow, for instance, pulmonary congestion, edema of the lungs, bronchitis, enlargement of the liver, and dropsy The circulatory background of these changes is indicated by hypertrophy of the left ventricle and increased tension of the arteries The latter become hard and tortuous

C THE IRREGULAR GOUT

We speak of gout as being irregular if it appears in places other than in the joints Types of irregular gout are seen frequently in persons who never have had any joint involvement, but who nevertheless are predisposed to gout, either by inheritance or by their habits of living It is important to be familiar with these types of gout, because one cannot depend for the diagnosis entirely upon the classical symptoms described by Sydenham

The signs of irregular gout usually are cramps and aching pains in the muscles, and tingling sensations in the hands and feet (Luff[9]) They are due to the precipitation of sodium biurate in crystalline form into the organs affected Sir William Roberts[1] mentions the deposition of these biurates in the viscera, he found them after death in the valves of the heart, the walls of the arteries and veins, in the vocal cords, in the mucous follicles of the pharynx, in the walls of the bronchial tubes, and even in the meninges

It is easy to see how difficult it is to recognize these cases of irregular gout In general, we can divide irregular gout into the following groups

1 Affection of the alimentary tract We speak of a gouty pharyngitis, gouty dyspepsia, or gouty intestinal catarrh

2 Cases involving the air passages and the lungs, such as gouty laryngitis, tracheitis, bronchitis, chronic gouty asthma, or gouty pulmonary congestion

3 Circulatory gout affecting the heart and vessles with anginal or pseudo anginal attacks and gouty phlebitis

4 Gout involving the nervous system, manifested by gouty migraine, neuralgia neuritis, etc

5 Irregular gout affecting the genito urinary system Here the most important feature is the gouty kidney, the inadequate function of which does not permit the proper elimination of uric acid These are cases which suffer from uric acid gravel and calculi, and not uncommonly have true gouty attacks later Also an irritability of the bladder may exist, with scanty urine and high specific gravity, and copious and large deposits of amorphous urates

6 The gouty eye One extra articular feature of gout to which attention has been called by Tuohy[14] is the so called "hot eye" This condition was first

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FIG. 78 Gout Note punched out areas at bases and heads of phalanges Case H D
FIG 79 Gouty bone cyst in head of midphalanx of fourth finger Case J E

cent The uric acid content of the blood of a known gouty patient varies according to the stage of the disease While a high uric acid level indicates gout, one should not make the diagnosis absolutely dependent upon this one feature

V THE X RAY SIGNS OF GOUT

The x ray changes are not always typical, but there are certain features which are suggestive of gout Particularly significant are small areas circular in outline, with very sharp borders, usually found in the epiphyses of the affected joint, especially of the toes and fingers They are due to absorption of calcium in areas in which sodium biurate has been deposited We see such semilunar cut-out effects especially in the metatarsal of the big toe They also appear as small indentations half or three-quarter circles, at the articular surfaces of the phalanges (Fig 78)



FIG 80 Extensive destruction of big toe joint in gout

In later stages there appears a cystic translucency of destruction at the joint often ending with complete obliteration of the articulation (Fig 79) In still later stages the distal ends of the metatarsals may become entirely destroyed so that the joint practically collapses In extreme cases, it comes to

described by Jonathan Hutchinson (1885) It affects both eyes, the conjunctiva becomes red, and the eyeballs feel hot and prickly as though irritated by sand It occurs usually within half an hour after a meal and may last a few hours or even a day Tuohy has followed a patient for over two weeks who had a uric acid level of 5 mgm per cent, and he was able to control the condition entirely by anti gout therapy

7 Finally, there is the gouty affection of the skin, which is manifested in form of an eczema, herpes, pruritis or urticaria

Irregular Gout

Case A P (Male)	#38 20732
Age 54 years	Adm November 5, 1944

This patient used alcohol freely He came with a year's history of pain in the feet in sudden attacks subsiding within a week He had suffered 18 to 20 of these attacks For the past three years he had suffered intermittent, generalized itching For two years he had a right trigeminal neuralgia characterized by sudden paroxysmal and severe pain in the face, set off by cold or any other stimulus Later the joint pain became more generalized involving the wrists, fingers shoulders and elbows The examination showed general excoriations of the skin, an enlarged olecranon bursa and draining sinuses from the dorsum of the foot There were albumin and granular casts in the urine, which led to the diagnosis of gouty nephritis Examination of the exudate from the foot showed urate crystals and calcium salts Blood examination showed a uric acid content of 7 7 to 9 mgm per cent

This case demonstrates a combination of true articular gout with gouty kidney, gouty affections of the skin, and a trigeminal neuralgia

IV THE DIAGNOSIS

A CLINICAL

So far as the diagnosis is concerned the most important clinical sign is the involvement of the big toe joint next, the ankle, knee, and the small joints of the hand and wrist In addition to this, gouty signs include swelling and hyperemia of the ligamentous tissues, effusion into the joint, and invasion of the capsule and other structures by uric acid deposits Tophi appear later in the cartilage of the ear and the nose, the eyelids, and even the larynx Most characteristic are the tophi in the cartilage of the ear, but it must be remembered that they do not appear early in the disease, and sometimes are not found for many years

B LABORATORY

The pathognomonic laboratory evidence is the high uric acid content of the blood The normal amount is 1 7 mgm per 100 cc with a certain latitude up to about 4 mgm which is a high normal In gout, this is very much increased, the average being 3 7 mgm per 100 cc of blood, but cases have been noted with as high as 8 mgm per cent and in one of our cases it reached 9 mgm per



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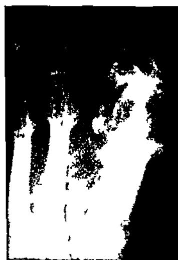


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FIG 81 Gouty destruction Case C C a) Elbow with large cavities in ulna b) Gouty cavities in humerus c) Gouty destruction of the knee joint

destruction of the entire metatarsal or metacarpal heads or the bases of the phalanges, with gross deformities and deviations (Fig 80) The x ray picture in other articulations, for instance, the elbow, shoulder, and spine, shows similar translucencies, erosions and marginal defects (Fig 81)

VI THE TREATMENT OF GOUT

A GENERAL HYGIENIC AND DIETARY REGIME

The treatment is principally hygienic It consists in temperate living, abstinence from alcohol, moderate eating, living in the open air, good elimination, good diet, warm clothing, and avoidance of rapid changes in temperature The restriction of diet concerns starchy or saccharine foods, but of particular importance is the elimination of food stuffs rich in purine, such as beef extract, beef broth, liver, kidney and brain Ebstein[3] urges the use of fat and butter and advises that gouty patients should, as far as possible, restrict the use of common salt Alcohol in any form is harmful and should be eliminated The use of mineral waters is supposed to have a beneficial effect on gout, although their value is doubtful Special usefulness is ascribed to waters which contain lithia, as in Saratoga and White Sulphur Springs in this country, Buxton and Bath in England, and Aix les Bains in France

B THE TREATMENT OF THE ACUTE ATTACK

The patient should be put to rest, with a mild diet and abundant fluids His limbs should be elevated wrapped in cotton, and he should have warm fomentation Opiates may become necessary where there is a great deal of restless ness and pain Locally, hot air treatment or passive hyperemia is advised The patient should be given energetic cathartics

The medication most widely used is wine or tincture of Colchicum which is given in doses of 20 to 30 grains every four hours with citrate of potassium

It has a powerful effect and promptly reduces pain and swelling The drug itself, Colchicum, must be given in very small doses, such as 0 01 grain every four hours, while the acute phase lasts We usually use wine or tincture of Colchicum

Equally effective but somewhat toxic is cinchophen or atophan It is useful in subacute attacks, in doses of 7 5 grains three to four times a day One should exercise care in the use of cinchophen because of its effect upon the kidneys and upon the production of white blood cells, which may become considerably restricted In case of a leukopenia, it must be discontinued

Vitamins as an adjuvant to the treatment have been introduced lately (Callahan and Ingham[2]) Nine cases were reported where the treatment con sisted in massive doses of vitamin B_1, or thiamin chloride, from 4,500 to 9,000 IU In addition to this, cinchophen was given in 7 5 grain doses three times daily, together with a purine free diet, warm mineral baths, and packs

C THE TREATMENT OF TOPHI

Sometimes it becomes necessary to deal with the tophi operatively This may be for cosmetic reasons, when they have become large and unsightly, for instance, in the subcutaneous tissues of the fingers, also, they may be painful, especially if located over exposed areas of the body, for instance, in the oleocranon bursa Furthermore, tophi may interfere with the movement of tendons of adjacent joints, or they may perforate through the skin and produce discharging sinuses A carefully planned operation will result in relief of pain and restoration of function In many instances, it is impossible to eradicate a tophus completely Before any surgical procedure on the tophi is planned, however, the patient's condition should be well fortified by administration of Colchicum or other anti gout medication, in order to insure control of the disease, because it is well known that surgical interference may easily precipitate a gouty attack

D CORRECTION OF GOUTY DEFORMITY

So far as the correction of a gouty deformity is concerned, it should be treated along the ordinary orthopedic principles As in all surgical procedures, a preliminary treatment with Colchicum or cinchophen is essential

REFERENCES

1 BEITZKE H *Ztschr f klin Med 74* 215 1912
2 CALLAHAN E J and INGHAM D W *Med Rec 149* 167 1939
3 EBSTEIN W *Die Natur und Behandlung der Gicht* Wiesb 1882
4 GARROD A B *Treatise on Gout and Rheumatic Gout* 1859
5 HEINE J *Virchows Arch f path Anat 260* 521 1926
6 HIPPOCRATES *Collected Works* Translated by R Fucks Munich H Luneberg *2* 363 1895
7 JAKSCH R v *Deutsche med Wchnschr 16* 741 1890
8 LANGE F J *Handb d Spez Pathol Anat u Histolog* Berlin O Lubarsch and F Henke *93* 309 1939



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Lecture VII

ON PYOGENIC ARTHRITIS

THE TERM indicates an inflammatory process of the synovial membrane and its adjacent structures caused by pyogenic bacteria We have met with pyogenic arthritis before, in the study of osteomyelitis, where the joint infection represented the extension of the osteomyelitic process into the articulation

The reason for referring to the subject now as a separate entity is because its relation to osteomyelitis represents only one aspect of the pathogenesis There are other sources of infection besides the extension from a neighboring osteomyelitic focus, pyogenic arthritis may be caused by direct implantation, it may also be a metastatic localization through the hematogenous route from a remote focus in the body In the latter case the systemic infection precedes the seeding of pyogenic bacteria in the synovial membrane, as it does in hematogenous osteomyelitis

I THE PATHOGENESIS

Any of the known pathogenic bacteria may be responsible for pyogenic arthritis The principal ones are the staphylococcus and the streptococcus The staphylococcus infection, as a rule, produces early and extensive suppuration, while the streptococcus infection usually shows an absence of suppuration and an absence of necrosis On the other hand, the streptococcus is more likely to cause secondary infection of the blood stream from a primary arthritis than is the staphylococcus Other infectious agents are the bacilli or cocci of diphtheria, cerebrospinal meningitis, pneumonia, typhoid, gonorrhea, and Malta fever

A THE DIRECT IMPLANTATION TYPE

The most frequent cause is external trauma, a compound fracture, or a gunshot wound Less common is the implantation of infectious material following operative procedures

The inflammatory reaction occurs within twelve hours after implantation The first hours after injury, therefore, are the most favorable for debriding and sterilizing the joint by operative interference The secondary systemic infection which follows a direct implantation type is not as severe as is the primary bacteremia in the hematogenous arthritis, and the condition is more likely to remain localized Nevertheless, there are numerous cases in which the local infection extends rapidly, and invades the system, causing a severe systemic septicemia As a matter of fact, a secondary systemic infection due to septic arthritis is more serious and more frequent than that following the implantation type of osteomyelitis

9 Luff A P *Gout, Its Pathology and Treatment* New York William Wood & Co 1899
10 Pommer G A *Microscop Unters d Gelenkgicht* Jena J Fischer, 1929 also *Wien Klin Wchnschr* *45* 801 and 845 1932 also *Beitr z Path Anat u z allg Path*, *90* 513 1933, and *95* 92, 1935
11 Price N L *Lancet 1* 22, 1939
12 Roberts Sir William *Uric Acid Gravel and Gout* Cronian Lecture 1892
13 Sydenham T *Tractatus de Podagra et Hydrope* London 1895 (Re edition)
14 Tuohy E L *Minnesota Med*, *24* 215, 1941

Lecture VII

ON PYOGENIC ARTHRITIS

THE TERM indicates an inflammatory process of the synovial membrane and its adjacent structures caused by pyogenic bacteria We have met with pyogenic arthritis before, in the study of osteomyelitis, where the joint infection represented the extension of the osteomyelitic process into the articulation

The reason for referring to the subject now as a separate entity is because its relation to osteomyelitis represents only one aspect of the pathogenesis There are other sources of infection besides the extension from a neighboring osteomyelitic focus, pyogenic arthritis may be caused by direct implantation, it may also be a metastatic localization through the hematogenous route from a remote focus in the body In the latter case the systemic infection precedes the seeding of pyogenic bacteria in the synovial membrane, as it does in hematogenous osteomyelitis

I THE PATHOGENESIS

Any of the known pathogenic bacteria may be responsible for pyogenic arthritis The principal ones are the staphylococcus and the streptococcus The staphylococcus infection, as a rule, produces early and extensive suppuration, while the streptococcus infection usually shows an absence of suppuration and an absence of necrosis On the other hand, the streptococcus is more likely to cause secondary infection of the blood stream from a primary arthritis than is the staphylococcus Other infectious agents are the bacilli or cocci of diphtheria, cerebrospinal meningitis, pneumonia, typhoid, gonorrhea, and Malta fever

A THE DIRECT IMPLANTATION TYPE

The most frequent cause is external trauma, a compound fracture, or a gunshot wound Less common is the implantation of infectious material following operative procedures

The inflammatory reaction occurs within twelve hours after implantation The first hours after injury, therefore, are the most favorable for debriding and sterilizing the joint by operative interference The secondary systemic infection which follows a direct implantation type is not as severe as is the primary bacteremia in the hematogenous arthritis, and the condition is more likely to remain localized Nevertheless, there are numerous cases in which the local infection extends rapidly, and invades the system, causing a severe systemic septicemia As a matter of fact, a secondary systemic infection due to septic arthritis is more serious and more frequent than that following the implantation type of osteomyelitis

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Fig 83 Suppurative arthritis Inflammatory cells are penetrating into and destroying the joint cartilage Inflammation granulation tissue and fibrosis of the marrow destruction of cartilage by proteolytic ferment

not only the synovial membrane but also the subsynovial tissues, the periarticular structures and the ligamentous apparatus in other words a true panarthritis

The intra articular abscess contains a proteolytic ferment, which together with the existing mechanical pressure causes the cartilage to become destroyed The destruction of the cartilage is further facilitated by the formation of a pannus formed from the infectious granulations of the synovial membrane and extending over the cartilage We find cartilage destruction most marked in the center of the joint where the joint bodies come into contact, it is less marked in the periphery This is in contrast to tuberculous osteomyelitis (Phemister[10]) (Fig 82)

This empyema of the joint consists of a yellowish exudate containing fibrin and detritus, and of the markedly injected synovial membrane (Chiari[3]) Microscopically, the latter contains a large number of mononuclear cells, and a heavy leukocytic infiltration can be found in the loose subsynovial layer The cartilage responds at first with a disappearance of its basophilic ground substance, but soon definite degenerative changes appear in the form of fibrillation and necrosis, produced by the action of the toxins and the proteolytic ferment (Fig 83)

In the panarthritis, or so called capsular phlegmon, we find that not only the synovial membrane but also the periarticular structures are infiltrated so that the subsynovial pus formation extends into the fibrous capsule The whole capsule may then be transformed into a jelly like infiltrated mass Finally, it comes to maceration and perforation of the capsule, and a periarticular abscess develops which spreads into the neighboring bursae and tendon sheaths

In the meantime, the destruction of the joint cartilage proceeds (Fig 84) Necrotic pieces of it may be found in the joint In the areas of denuded cartilage the subcortical bone is exposed (Fig 85) Then follows the destruction of the subcortical bone which may lead to pathological dislocation, facilitated by the relaxation or perforation of the capsular apparatus

The reparative stage Reparative changes are initiated by the transformation of the infectious granulation tissue into fibrous tissue This tissue sets up a barrier against the deep penetration of the infection, by means of fibrous proliferation of the subchondral marrow which is produced by the endosteum and

B THE EXTENSION TYPE

In a certain sense, this is also an implantation type except that the infection does not come from the outside but from neighboring tissues A frequent source is the osteomyelitis of the adjacent bones, the primary focus being located in the metaphysis or epiphysis In a series reported by Kulowski,[9] 21 per cent of all cases of osteomyelitis of the long bones sooner or later developed suppurative *arthritis of the neighboring joints by extension of the osteomyelitis focus*

C THE HEMATOGENOUS OR EMBOLIC TYPE OF PYOGENIC ARTHRITIS

The infection occurs through the blood stream from a primary focus situated in a remote part of the body This is the most common type The primary focus may be a furuncle or an abscess, a bursitis, a puerperal sepsis or an otitis media, or it may be a general infectious disease, particularly measles and scarlet fever

II THE PATHOLOGY

The pathological events more or less follow in a certain sequence, but there is some difference in the pathological development according to the invading organism

A THE STAPHYLOCOCCUS INFECTION

In *the initial stage,* we notice an increase in the hydrostatic pressure due to serous effusion The pressure blocks the lymphatic absorption, it also has a mechanical effect, because it extends and stretches the capsular reinforcement and produces instability of the joint

The serous stage Nearly all infection begins with a serous exudate coming from the hyperemic synovial membrane This exudate contains mucin, leukocytes, and flakes of fibrin The cartilage up to this stage remains unchanged, and the viscosity of the joint fluid is increased



Fig 87 Suppurative arthritis central destruction of cartilage Case D A

Then follows the *seropurulent stage* Here we already find excessive damage to the synovia, and the joint fluid appears milky and finally becomes purulent The periarticular structures become invaded and water logged, as the relaxation of these structures increases, it may finally lead to subluxation or even dislocation of the joint

Finally, there is the *stage of suppuration,* in which the contents of the joint consist entirely of pus, a joint empyema All structures are involved,

not only the synovial membrane but also the subsynovial tissues, the periarticular structures, and the ligamentous apparatus, in other words, a true panarthritis

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Fig 84 (Left) Suppurative arthritis Joint cartilage has been completely destroyed inflammatory cells are seen in the marrow and over the subchondral plate

Fig 85 (Right) Old suppurative arthritis Joint cartilage has been almost completely destroyed *vascular granulation tissue is seen in the marrow spaces perforating the subchondral plate* exposure of subchondral bone Case M P

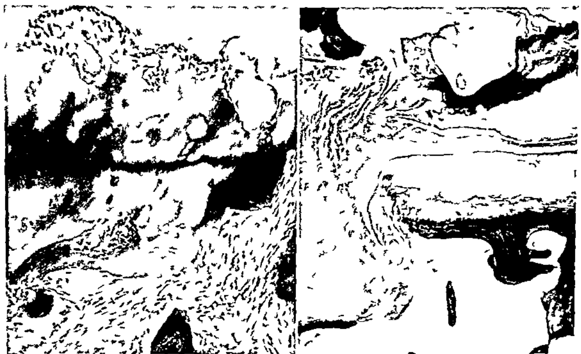


Fig 86 (Left) Old suppurative arthritis The bone is necrotic the suppurative process has been followed by organization with formation of young granulation tissue and scar tissue The joint cartilage is necrotic and has been reduced in thickness down even to the calcified layer Granulation tissue is seen covering the joint surface and merging with the fibrotic bone marrow Case M P

Fig 87 (Right) Suppurative arthritis of the subastragalar joint with fibrous ankylosis the cartilaginous surfaces are separated from each other by a thin strand of fibrous pannus Fibrous tissue has already entirely replaced the cartilage and a fibrous ankylosis exists the bone trabeculae are porotic Case C B

eventually separates the necrotic area from the living bone (Fig 86) The bone marrow tongues which invade the granulation tissue from the subchondral bone may carry enough osteogenetic power to produce bone formation in this scar tissue In many cases, however, the end product of this regenerative process remains as a tough, fibrous scar which fills the entire joint The defects caused by the ulcerated cartilage are closed up by granulation tissue coming from both the pannus of the synovial membrane and from the endosteum of the epiphysis (Fig 87) Even though there occurs no metaplastic bone formation by bone marrow tongues streaming into the fibrous scar, the fibrosis of the bone marrow in the cancellous bone itself leads to a reactive bone formation, and the subchondral bone becomes sclerotic, somewhat like that seen in arthritis deformans

When bony ankylosis of the joint is produced by bony bridging and by firm osseous union, one observes that in the course of time a static rearrangement of the architecture of the bone takes place, so that the bone trabeculae are actually crossing the joint without interruption But this is a very slow procedure and can be observed only in old septic joints, which have resulted in complete bony ankylosis

B THE STREPTOCOCCUS INFECTION

The pathological events in the case of of streptococcus infection are somewhat different This infection is usually secondary to another focus in the body, such as middle ear infection or tonsillitis, scarlet fever or erysipelas Scarlet fever is a frequent offender in causing metastatic joint infection

A characteristic of streptococcus arthritis is the serous type of exudate, and the tendency to pluriarticular involvement The aspiration of the joint yields streptococcus, but occasionally the culture may remain sterile Eventually, the streptococcus arthritis leads to suppuration, usually after the third or fourth week, so that one may distinguish a serous and a purulent phase In the purulent phase, the infection spreads rapidly through the synovia into the periarticular tissues It leads to loosening of the capsular apparatus and to abnormal mobility of the joint It is in this phase that the cartilage succumbs in large placques, and the subchondral bone becomes exposed extensively The final result is the destruction of the capsule and, not infrequently, a pathological dislocation

C THE PNEUMOCOCCUS

The pneumococcus arthritis, due to the pneumococcus of Frankel Weichselbaum was first described by Grisolle (1884) and Schuller (1885) [13] Sometimes joint symptoms occur very early in the disease, even preceding those of the lung More frequently, however the joints become involved during convalescence The lungs are not the only port of entry, often a pneumococcus otitis media or tonsillitis is the primary focus The monarthritic form is prevalent on

the whole, although the shoulders, knees, elbow, sternoclavicular joints, wrists and ankles may become affected together

Here, also, a serous and a purulent phase can be distinguished but, in contrast to the streptococcus infection, the serous form is less common than the purulent one, which produces a thick creamy pus The capsule shows phlegmonous infiltration, and the synovial membrane appears coated with fibrin containing displococci The destruction of cartilage is not as extensive, and the end effect is usually regeneration Some of the milder cases heal without residuals

D THE MENINGOCOCCUS

The meningococcus or Weichselbaum's diplococcus is occasionally the cause of purulent arthritis, appearing as a sequela to meningitis Joint infection occurs usually four to six days after the onset, and its frequency in meningitis is 0 6 per cent in infants, 4 4 per cent in children, but as high as 16 per cent in adults (Sainton and Maille[1])

The monarticular form prevails Knees, ankles, sternoclavicular joints, hips, and wrists are favored locations The pathological changes are those already described, a pyarthrosis with a fibropurulent, light greenish exudate and with hyperemia, swelling and edema of the synovial membrane The microscope shows intense leukocytic infiltration and many meningococci Regeneration to normal also occurs not infrequently without subsequent ankylosis

III THE CLINICAL PATHOLOGY

A THE GENERAL SYMPTOMS OF PYOGENIC ARTHRITIS

In the vast majority of cases, the onset in the hematogenous type is acute and accompanied by marked constitutional reaction in the form of chills, high temperature, and protration In the implantation type the general symptoms are not prominent, since septicemia usually does not occur

B THE LOCAL SIGNS

In the hematogenous type some time may elapse before local signs develop In the direct contamination type the local signs begin after a few hours The principal local signs are pain local tenderness, redness and later, fluctuation and edema restriction of motion and contracture This is accompanied by rapid atrophy of the muscles

1 The pain Pain in most cases is spontaneous There is definite tenderness to pressure, usually corresponding to the lines of synovial reflection

2 Motion Motion is always painful and is resisted by spastic contracture of the muscles The joint is held in midposition Restriction of motion is concentric, that is, equal in all directions

3 Redness and heat are more apparent in the more superficially situated

joints, such as the knee or the wrist It is neither an early nor a constant sign in more deeply situated joints, such as the hip or the shoulder

4 Swelling and fluctuation occur with an increase of the exudate The swelling is accompanied by edema of the dependent portions of the limb

5 Muscular atrophy develops rapidly, it is noticed especially in the quadriceps when the knee joint, and in the deltoid when the shoulder joint is involved

C THE LATER SIGNS AND DEFORMITIES

Deformities are of three types Faulty position, ankylosis, and loss of length The greatest amount of disability occurs where massive osteomyelitis has led to infection of the joint, and a considerable amount of bone has been destroyed Cases in which relaxation or perforation of the capsule has taken place during the acute stage often end up with pathological dislocations, this sometimes occurs before there is extensive destruction of the joint itself, especially in very young children

IV THE X RAY FINDINGS

Unless there is a neighboring osteomyelitic focus penetrating into the joint, the x ray evidence of pyogenic arthritis develops slowly The first sign is the capsular distention, recognizable by the bulging shadow of the joint capsule Next, there is the regional bone atrophy Then one sees a narrowing of the joint space due to degeneration and destruction of the cartilage (Fig 88) Following this, erosions are seen, first at the contact points of the joint constituents They indicate that the cartilage has become completely worn off and that the subchondral layer of bone has been laid bare The later x ray picture shows the progressive destruction of the joint The joint outlines become serrated, irregular, and punched out, and finally signs of extensive loss of bone are noted As this destruction goes on, entire portions of the joint may disappear The femoral head may become com



Fig 88 Pyogenic arthritis narrowing of the joint space

pletely absorbed, and the acetabulum may be greatly enlarged by erosion Some cases will end in fibrous union, others, in complete ankylosis, and still others, in pathological dislocation The final x ray varies with the definite end result of the infection (Fig 89)

V THE LABORATORY FINDINGS

The diagnosis can be made by aspirating the joint and examining the exudate The joint fluid contains a high amount of leukocytes with a preponderance of polymorphonuclears

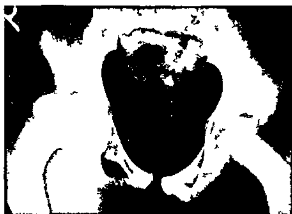


Fig 89 Suppurative arthritis of the right hip beginning dislocation

A blood culture should be taken in every case During the acute infection the blood will show considerable leukocytosis, which may reach 30 or 40 thousand In the chronic stage a moderate anemia may be noticed The sedimentation time is of importance, it is always accelerated in the acute phases The urine must be examined for signs of kidney damage, particularly in protracted cases which are apt to produce amyloidosis of the parenchymatous organs

VI THE DIFFERENTIAL DIAGNOSIS

A RHEUMATIC FEVER

This condition is polyarticular It responds to salicylates, and it does not result in suppuration

B TUBERCULOSIS

The differentiation here is more difficult, because both pyogenic arthritis and tuberculosis are usually monarticular However, the course of tuberculosis is eminently chronic and, as the x ray shows, the destruction of the joint is marginal, and there is no sclerosis

C BRUCELLOSIS

Malta fever infection of the joint usually takes a more chronic course It rarely comes to suppuration, and the blood examination often shows a low white count The diagnosis is established by the positive agglutination test (Fig 90)

VII THE TREATMENT OF PYOGENIC ARTHRITIS

A THE TREATMENT OF THE FOCUS OF INFECTION

An infected tonsil, a purulent sinus or, more often, an otitis media may still be active when the metastatic focus in the joint has made its appearance In

this event, evacuation of the joint should be the first consideration Drainage or aspiration must not be postponed until after removal of the focus, but it should be immediate

B THE SUPPORTIVE GENERAL TREATMENT

Supportive treatment is just as essential in pyogenic arthritis as in osteomyelitis, and there should be no delay in carrying it out It consists in the usual administration of fluids, in blood transfusion, in rest, and especially in use of

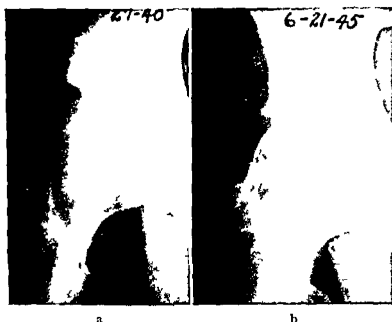


Fig 90 Brucellosis of the hip joint Case W R (male), #40 15851, 28 years, November 1940 Involvement of right hip for four months following general malaise, hip is red and tender Agglutination test positive for Malta fever X ray shows narrowing of the joint space a focus in the acetabulum and destruction of the joint cartilage There was an abscess of the right thigh below Poupart's ligament with a draining sinus culture was positive for Brucella suis Conservative treatment resulted in ankylosis of the hip in 1945 a) November 1940 b) Ankylosis in June, 1945

sulfonamides and penicillin We recommend sulfathiazole for the staphylococcus, and sulfadiazine and sulfanilamide for the streptococcus infection The preference today is for penicillin, which should be given at once, as soon as a test for penicillin resistance has been made The dosage and technique are the same as for osteomyelitis

But one should not rely on sulfonamides or penicillin alone in the treatment of septic arthritis The joint must be evacuated In the streptococcus infection the aspiration performed to identify the bacteria may be sufficient under the protection of penicillin and sulfonamides, provided the limb is strictly immobilized In cases of pyogenic arthritis due to staphylococcus, however, drainage should be carried out at the earliest possible moment We have seen too many cases in which the joint was destroyed because of delayed drainage (Fig 91)

It has been remarked very appropriately by Kulowski[9] that use of the anti-

biotics increases the surgeon's responsibility due to the fact that it, so to speak, throws a mask over the local lesion The possibility that the quiescence of the joint may only be apparent and that symptoms may reappear soon after the drug is discontinued should always be kept in mind Sulfonamides and penicillin are bacteriostatic agents which merely check and localize the joint infection, the actual sterilization of the joint is accomplished by the defensive forces of the body

C THE LOCAL TREATMENT

1 Evacuation of the joint

INDICATIONS BY TYPE OF INFECTION

Joint evacuation should be carried out immediately It must be extensive enough in staphylococcus infection to allow rapid evacuation of the exudate It can be confined to simple aspiration in streptococcus infection, where it merely lessens the intra articular pressure and makes absorption of the contents possible In either case, the joint must be immobilized in order to limit the spread of infection and provide the resistance necessary for its control, and to prevent subsequent deformity

INDICATIONS BY TYPE OF EXUDATE

In the serous type, which is usually a streptococcus infection, aspiration together with immobilization is sufficient One must first aspirate, and if staphylococcus is found the incision must be made promptly and freely The purulent type requires immediate and adequate drainage with counter drainage if necessary Immobilization in plaster or by traction or brace must follow in any event

TECHNICAL PRINCIPLES OF JOINT EVACUATION DRAINAGE

The principles of Orr are wide drainage without closure, and adequate immobilization The adequacy of drainage is shown by the rapid recession of inflammatory signs and the favorable general reaction Secondary closure can be carried out as soon as the wound appears sufficiently clean and healthy

Resection In some cases of excessive suppuration, soft tissue drainage is inadequate, and joint resection is required This applies particularly to the hip joint, where recesses favor the accumulation of pus Resection is also necessary in cases of extensive necrosis or sequestration, for which simple drainage would not be adequate

2 Joint fusion

The natural outcome of the septic process is a fibrous ankylosis, except in those cases in which the joint has been saved by early drainage In more virulent types, the resulting ankylosis may be bony The tendency to spontaneous bony fusion is much greater in the pyogenic than in the tuberculous joint

When the end result is a fibrous ankylosis with more or less instability and pain on weight bearing, a fusion operation is indicated The time for ankylosing operations on septic joints is the period of absolute quiescence The decision is made on the basis of (a) the local condition (no signs of inflammatory activity and no increase of contractures) and (b) the x ray (sharpening of the joint contours and sclerosing reactions around areas of destruction)

3 Joint mobilization

So far as the prevention of contracture is concerned, it is a matter of judgment how soon during the disease careful mobilizing maneuvers can be instituted It is a safe rule not to begin until the patient himself can perform them actively Even then, these attempts should be made carefully and should be checked closely, because they may cause an accentuation of the inflammatory condition During the first world war, active mobilization of the knee joint was advocated under the so called Willems' treatment, motion being instituted even before the drainage had ceased After adequate drainage, the patient was encouraged to carry out active motion This seems a contradiction to the principle of absolute rest and immobilization which we have so strongly emphasized in the early treatment Though it is possible that a certain amount of active movement will be tolerated without exacerbation of the infectious process, on the whole it is a dangerous procedure which must be strictly supervised if it is established at all Good judgment based on daily observation is required to determine just how much active motion a joint can carry out under these circumstances

Nothing is gained by trying to overcome malposition of a joint by forcible *manipulation, it is a procedure which cannot be condemned too strongly* While it is possible during the chronic or subacute stage to accomplish a certain amount of gradual correction, by hinged braces or turnbuckles, any forcible manipulation should be avoided Even the gradual correction by the above-mentioned means must be closely supervised, as any attempt which goes beyond the tolerance of the joint is sure to result in reactivation of the inflammation

When the period of absolute quiescence has been reached, the question arises whether mobility can then be restored operatively by means of arthroplasty There are only four joints which lend themselves to this procedure at all, the hip joint, the elbow, the knee, and the metacarpophalangeal articulations If both hip joints are ankylosed following a septic arthritis, arthroplasty should be carried out in one hip, provided the joint is absolutely quiescent In the knee joint an arthroplasty should be performed also, if both knee joints are ankylosed or if one is ankylosed in unsuitable position, for instance, in complete extension or extreme flexion In the elbow joint the arthroplasty is indicated when both joints heal in complete extension, or when one joint heals in extension and the other in flexion, the joint in extension must be operated

The septic joint which results in bony ankylosis is much better suited for arthroplastic operations than any other type of destructive arthritis The

operation, however, requires not only complete clinical quiescence, but the x ray picture must also show that the static realignment of the bone has already taken place and that the textural design of the bone blends intimately from one joint constituent into the other

Our statistics on arthroplasty in septic joints are as follows [7] The cases of ankylosis following suppurative arthritis of the hip joint[5] show a relatively high rate of good results (44 4 per cent) In arthroplasty of the elbow joint for ankylosed septic arthritis[7] the results were 33 per cent excellent and 33 per cent good The arthroplasty of the knee joint showed 33 per cent good results Our over all results in arthroplasties performed for suppurative arthritis are 53 5 per cent good, 21 4 per cent fair, and 25 per cent poor, for all joints

4 **The operative indications for deformity**

Ankylosis in faulty position is corrected by osteotomy, but it should be performed always extra articularly, that is, some distance from the infected joint It, too, requires complete quiescence, otherwise, there is likely to be a recurrence of the old infection Looking over our series of cases (J B Davis[4]), we found that if an osteotomy for correction of the deformity was performed through the old diseased area, the percentage of recurrence was 31 5 per cent, even though there was an average period of quiscence of 4 4 years On the other hand, when the operation was carried out near but not through the old seat of disease, the percentage of recurrence dropped to 17 6 This indicates that the time interval is of less importance than is the selection of the site for osteotomy

VIII THE SPECIAL CLINICAL PATHOLOGY OF THE SUPPURATIVE ARTHRITIS

A THE SACRO ILIAC JOINT

Among our 30 cases of pyogenic arthritis of the sacro iliac joint, 8 were due to extension from the lumbar spine Whether they are primary or secondary infections, the treatment is the same, namely, radical resection and drainage The method we use with preference is that of Picque, which consists in resection of the posterior wing of the os ilii and adjacent portions of the sacrum

B THE HIP JOINT

The greatest amount of destruction is produced by the staphylococcus, especially in young children Only by immediate and adequate drainage can one save the head, in contrast to the situation in streptococcus and pneumococcus infections which cause much less destruction and for which a simple aspiration may suffice Unless adequate drainage is performed, the entire head may succumb and may remain as a sequestrum in the joint (Fig 91)

Another frequent complication of suppurative arthritis of the hip joint is the pathological dislocation *It is due either to early perforation of the capsular sac*

or to extensive destruction of the acetabulum and upward displacement of the head Three types of pathological dislocations can be distinguished The type in which the head dislocates because of early perforation of the capsule is seen in very young infants There is very little destruction of the head because the early perforation of the capsular sac lets the head escape and saves it from destruction *In the second group the head remains* in the capsular sac, but the acetabulum becomes destroyed, and the dislocation is secondary to the acetabular destruction (Fig 92) In the third, the most common group, *both the acetabulum and the head, and even* the major portion of the neck are destroyed The degree of destruction of the head and acetabulum is commensurate with the amount of time which has elapsed between the appearance of symptoms and adequate drainage (Fig 93)



Fig 91 Pyogenic *arthritis of the hip* joint loss of the head because of delayed drainage Case B P

To obtain adequate drainage it may be necessary in extreme cases to resort to resection of the joint *Included here are those cases in which an actual* sequestration of the head has taken place and in which complete drainage cannot otherwise be obtained

Of 19 cases of pyogenic arthritis of the hip joint seen during 1937-47, four resulted in complete, painless motion after adequate drainage was performed *one to four weeks after the onset One case had 75 per cent painless motion*



Fig 92 (Left) Septic arthritis pathological dislocation *of the hip* Case C W *(female)* #40 6859 8 years May 1940 Infection of the right hip of 10 months duration abscess and sinus formation and no adequate drainage Complication of osteomyelitis of the left and right tibiae *and laminae* of second and third lumbar X ray shows subluxation of the right hip following purulent arthritis enlarged acetabulum Drainage of tibiae lumbar spine and right hip healing of hip in faulty position Observation seven years

Fig 93 (Right) Pyogenic arthritis of the hip

after early adequate drainage One case of low grade infection in a 54 year old man had only 35 per cent painless motion On the other hand, five cases ended in fibrous fusion, and all had either inadequate or late drainage Six cases had bony fusion, of which four had no drainage at all, one had late drainage, and one had inadequate drainage Two cases died These 19 cases clearly demonstrate the urgent necessity of adequate and early drainage [11]

According to G H Harmon and C O Adams,[8] the natural outcome in suppurative arthritis under the principles of treatment outlined above is as follows In infants up to two years, where the streptococcus infection is more prevalent, the head and neck of the femur become destroyed if drainage is not done immediately, or if early aspiration is not performed Otherwise, destruction of the head and pathological dislocation occur Infants and children have a tendency for rapid healing in streptococcus infection (Badgley[1]) Between the ages of three and five, there is a preponderance of the staphylococcus infection at ages up to 12 staphylococcus and streptococcus infection occurs with equal frequency, but here again the tendency to restriction of motion and to the formation of an ankylosis is greater In the age group from six to 18 years the staphylococcus prevails, according to Harmon and Adams,[8] and in many cases pathological dislocations with severe bone changes and ankylosis can be seen Of all the age groups, the infants who survive infection make the most rapid and most complete recovery, without sinuses and recurrences

C THE KNEE JOINT

In purulent infection, the knee joint goes into a pathological position of about 20 to 30 degrees flexion The joint fills up, becomes rapidly distended,

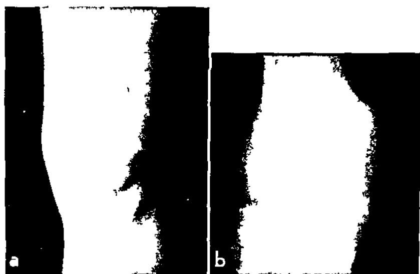


Fig 94 Septic arthritis of the knee aspiration Case J A (male) #21919 ?5 years December 1930 Acute left knee symptoms for two months fever of 105 degrees chills and prostration Aspiration and conservative treatment resulted in recovery with good motion Infection was presumably streptococcus but joint fluid culture was negative a) Joint swelling b) Narrowed joint space

and a relaxation of the capsular apparatus develops There is a tendency to tibial subluxation and valgus deformity, with external rotation of the tibia Streptococcus infection may be controlled by aspiration (Fig 94) Staphylococcus infection, which is more frequent, requires drainage according to the same principles applied to the hip The drainage is carried out by the parapatellar route, using the Orr technique

The penetrating wounds of the knee may be divided into cases in which there is no damage to the bone and cartilage, cases in which there is moderate damage to the condyles of the femur, and cases in which there is severe destruction of the joint components The routine treatment for these cases is the arthrotomy, the intra articular debridement, lavage and immobilization in a hip spica

The closure of the synovial membrane with immobilization is recommended in all cases in which there is either not any or only minimal damage to bone and cartilage This, however, is permissible only if the surgical treatment is instituted within eight to twelve hours after the wound has occurred If more time has elapsed, the closure of the synovial membrane should be omitted in order to permit drainage In all cases where there is severe damage to the bone, the wound should always be left open for drainage When the knee is hopelessly destroyed at the time of injury, resection will prevent chronic sepsis and promote early healing

D THE ANKLE JOINT

The acute swelling of the suppurating ankle joint obliterates the anatomical landmarks The foot is usually in equinus position The treatment is immediate drainage carried out from an anterior or anterolateral incision followed by application of a plaster cast which holds the foot in a favorable position, that is, neutral rotation and no more than 10 to 20 degrees plantar flexion When the destruction is extensive, however, this approach is inadequate and more radical measures are in order For very extensive suppuration, one may have to carry out an astragalectomy to establish adequate drainage If the purulent arthritis of the ankle joint is caused by osteomyelitis of a tarsal bone perforating into the joint, one will find that the other bones of the tarsus become involved very rapidly unless drainage is promptly provided

E THE FOOT

The osteomyelitis of the tarsal bones very often penetrates into the various articulations of the foot It is in this situation where drainage is especially important, because the infection makes rapid progress and, gaining the fascial planes and interstices, becomes uncontrollable One should always try to keep to the dorsal side in draining the joints, although the subastragaloid joint has to be drained from both sides On the whole, one should avoid plantar incisions

after early adequate drainage One case of low grade infection in a 54 year old man had only 35 per cent painless motion On the other hand, five cases ended in fibrous fusion, and all had either inadequate or late drainage Six cases had bony fusion, of which four had no drainage at all, one had late drainage, and one had inadequate drainage Two cases died These 19 cases clearly demonstrate the urgent necessity of adequate and early drainage [11]

According to G H Harmon and C O Adams,[8] the natural outcome in suppurative arthritis under the principles of treatment outlined above is as follows In infants up to two years, where the streptococcus infection is more prevalent, the head and neck of the femur become destroyed if drainage is not done immediately, or if early aspiration is not performed Otherwise, destruction of the head and pathological dislocation occur Infants and children have a tendency for rapid healing in streptococcus infection (Badgley[1]) Between the ages of three and five, there is a preponderance of the staphylococcus infection at ages up to 12 staphylococcus and streptococcus infection occurs with equal frequency, but here again the tendency to restriction of motion and to the formation of an ankylosis is greater In the age group from six to 18 years the staphylococcus prevails, according to Harmon and Adams,[8] and in many cases pathological dislocations with severe bone changes and ankylosis can be seen Of all the age groups, the infants who survive infection make the most rapid and most complete recovery, without sinuses and recurrences

C THE KNEE JOINT

In purulent infection, the knee joint goes into a pathological position of about 20 to 30 degrees flexion The joint fills up, becomes rapidly distended,

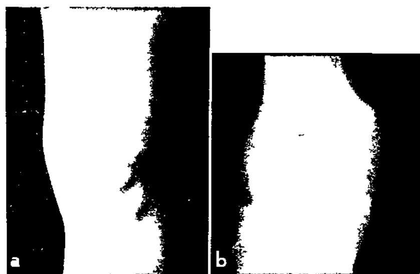


Fig 94 Septic arthritis of the knee aspiration Case J A (male) #21919 25 years December 1930 Acute left knee symptoms for two months fever of 103 degrees chills and prostration Aspiration and conservative treatment resulted in recovery with good motion Infection was presumably streptococcus but joint fluid culture was negative a) Joint swelling b) Narrowed joint space

and a relaxation of the capsular apparatus develops There is a tendency to tibial subluxation and valgus deformity, with external rotation of the tibia Streptococcus infection may be controlled by aspiration (Fig 94) Staphylococcus infection, which is more frequent, requires drainage according to the same principles applied to the hip The drainage is carried out by the parapatellar route, using the Orr technique

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F THE SHOULDER JOINT

As a rule, the destruction due to purulent arthritis is not as extensive in the shoulder as in the hip joint, but there is, on the other hand, a tendency to subluxation and to contracture For this reason, one must provide proper splinting after adequate drainage has been performed A resection of the head is necessary only in cases of extensive destruction Drainage can be carried out



Fig 95 Septic arthritis of the shoulder joint drainage Case E V (male) #42 1?619 55 years November 194? Swelling and pain of both shoulders for five months Both shoulders were drained the left healed with normal motion but the right remained swollen and continued draining X ray showed extensive destruction of the right humerus Extensive operative drainage was performed and penicillin given Culture showed hemolytic staphylococcus



Fig 96 Septic arthritis of the metacarpophalangeal joint ankylosis Case O S (female) #E 8360 26 years October 1930 Painful swelling of wrists following tooth extraction five weeks ago Incision and drainage of right wrist then involvement of metacarpophalangeal joint of left middle finger but no drainage was performed Examination showed ankylosed metacarpophalangeal joint of the left middle finger X ray destruction of the finger joint The right wrist was normal Finally drainage of the metacarpophalangeal joint was done Case demonstrates advantage of early drainage (right wrist) and disadvantage of delayed drainage (ankylosis of metacarpophalangeal joint)

by proceeding through the deltopectoral groove, and one should always establish posterior counter drainage, extreme cases may necessitate the adding of dependent drainage through the axilla (Fig 95)

G THE ELBOW JOINT

In draining the elbow joint, one should first expose the radiohumeral articulation because it is the easiest to approach If this approach is not sufficient, a second incision should be made for the drainage of the ulnohumeral portion at the inner side or posteromedially

H THE HAND AND WRIST

We prefer the dorsal approach for drainage If the tendon sheaths on the volar side are involved, they will have to be drained separately, according to the principles which govern the drainage of the radial and ulnar bursae They

should be opened along incisions which follow the radial side of the thumb and the ulnar side of the little finger On the other hand, the deep spaces, that is, the palmar and thenar spaces, require a special incision along the web between the thumb and index finger, or between the third and fourth fingers, or along the thenar and palmar creases The space of Perona may be reached between the deep flexors and the pronators quadratus at the forearm This space is often secondarily involved from a perforated purulent ulnar or radial bursa (Fig 96)

REFERENCES

1 Badgley, C E Iglesias L, Perhan W S and Snyder C H *J Bone & Joint Surg*, *18* 1047, 1936
2 Browne H G *Research Seminar Notes*, Dept Orthop Surg State Univ of Iowa *11 D* 49 1937
3 Chiari H *Handb d Spez Pathol Anat u Histolog*, Berlin O Lubarsch and F Henke *92* 12 1934
4 Davis J B *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa, *12 D* 21 1938
5 Fuiks D N *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *10 D* 89 1936
6 Hampton O P *J Bone & Joint Surg* *28* 659 1946
7 Hanavan E J *Research Seminar Notes*, Dept Orthop Surg State Univ of Iowa *19 D* 13 1948
8 Harmon G H and Adams C O *Surg Gynec & Obst* *78* 4 1944
9 Kulowski J Quoted by Ghormley R K *Orthopedic Surgery* New York Thomas Nelson & Sons 1938 p 50
10 Phemister D B *Proc Inst Med Chicago* *7* 169 1929
11 Picaso G *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *16 D* 13 1943 44
12 Sainton F and Maille J *Presse Med* *23* 345 1915
13 Schuller M *Arch f klin Chir* *31* 276 1885

Lecture VIII

ON SYPHILIS OF BONE AND JOINT

THE STORY of syphilis of the skeletal system is of great historical interest Old Indian skeletons show definite evidence that syphilitic lesions occurred long before the disease itself was recognized or even recorded We learn from William's account of the history of bone syphilis that no bones which are definitely syphilitic have been found in Europe before the voyage of Columbus to America, but that there is abundant evidence of bone syphilis antedating Columbus's voyage in the skeletal remains of the American Indians The so called Pecos cave of New Mexico contained a number of such skeletons, undoubtedly of pre Columbus date, probably 800 to 1,000 years old The characteristic lesions of the frontal bone which presented roughened surfaces and many low and flat elevations and depressions are now definitely recognized as syphilis

I THE CONGENITAL SYPHILIS OF BONE

A THE PATHOLOGY

Congenital syphilis of the bone presents itself in two rather distinct forms, according to whether it is manifested in fetal life and early infancy, or in the later life of the child The later manifestations correspond largely to those of the tertiary stage of the acquired syphilis, whereas the early congenital *lesions* seem to present a picture of their own During fetal life only the cartilaginous preformed bones are involved, and the lesions are usually symmetrical and multiple The membranous preformed bones, such as the flat bones and the skull *become involved after birth Only in second infancy does the lesion* develop a certain tendency to localize

1 The early hereditary syphilis of bone

There are three principal features characterizing this condition The first is the osteochondritis, discovered and described by Wegner in 1870, the second is the fetal type of syphilitic periostitis, first mentioned by Parrot[5] in 1872, the third is the diaphyseal osteomyelitis

Osteochondritis Here again three phases can be distinguished The first phase represents a broadened and irregularly outlined provisory calcification zone which has the aspect of a barbed wire formation In this zone, there appears a yellow or gray soft granulation tissue which extends into the cartilage and may eventually cause a separation of the epiphysis from the metaphysis To this spirochete infection the osteogenic tissue reacts with the arrest of its osteoblastic activity We see, therefore that the cartilage begins to proliferate

irregularly, and the cartilage columns become lengthened so that the whole calcification zone pulls out like a telescope (Fig 97) The granulation tissue represents the reaction of cartilage and bone marrow to the toxins of the spirochetes In a later stage of the syphilitic osteochondritis, the granulation tissue increases considerably It contains all kinds of cells, spindle shaped, perivascular accumulations, lymphoid infiltrations, and polynuclear cells, as well as many vessels Still later, regressive changes take place in this granulation tissue, such as necrosis or fat degeneration In time this granulation tissue completely interrupts the connection between epiphysis and metaphysis, and in so doing it may split the provisory calcification zone itself into an epiphyseal and a diaphyseal portion (Fig 98) The x ray then shows a more or less transparent zone, bordered by two narrow white bands, which represent these two calcification layers separated by a layer of granulation tissue (Fig 99)



FIG 97 Luetic osteochondritis Separation of the epiphysis granulation tissue growing into the cartilage pulled out calcification zone of cartilage

The x ray of the syphilitic osteochondritis is especially well seen at the lower end of the femur and the upper end of the tibia, also at the lower end of the radius and ulna, at the distal end of the metatarsals and metacarpals, and the proximal end of the phalanges The zone of increased calcification appears as a broad white band in which ossification is retarded Beyond this, the destruction of bone by granulation tissue is manifested by a zone of atrophy or absorption This is called the submetaphyseal rarefaction, it is due to the retardation of the osteoblastic activity by the infection (Fig 100) Under antiluetic treatment, the gran-



FIG 98 Newborn syphilitic baby Round cell infiltration and fibrosis in epiphysis invasion of the zone of cartilage proliferation between the columnizing and resting cartilage





FIG 100 Osteochondritis of the forearm bones Note absorption bands in metaphysis Case R W

FIG 99 Luetic osteochondritis of the newborn Case R P (male) #40 127 six weeks January 1940 Positive Wassermann crusts in nose ulcerations in mouth desquamation of skin X ray showed luetic octeochondritis Patient died soon after of pneumonia

ulation tissue becomes rapidly replaced by normal marrow, and the normal process of calcification is resumed However, the x ray picture still shows a wide band of rarefaction at the metaphysis

The separation of the epiphysis, first described by Parrot,[5] represents an end effect produced probably by mechanical movement of the fetus in utero Yet it cannot be considered as purely traumatic, because it is due to the poor cohesion between the metaphysis and epiphysis, which are held together only by granulation tissue so that shifting movements suffice to bring about the separation

PERIOSTITIS

This condition was first discovered and described also by Parrot,[5] in 1872 It is a specific lesion characteristic of early syphilis, although a secondary periostitis occurs later in infancy in connection with the repair of luetic osteitis (Fig 101) As a symptom of early congenital lues early periostitis is found in the first weeks of life in the surviving children, it is often combined with the above described osteochondritis, although the two processes may not run in equal intensity (Fig 102)

In the x ray picture, the periostitis appears along the long bones in the form of onion skin layers, often three or four layers piled upon each other These



Fig 101 Syphilitic osteoperiostiti (Slide from rib biopsy low power focus) Note periosteal new bone al o ubperiosteal and ubcortical granulation tissue



Fig 10? Syphilitic osteoperiostitis (Slide from rib biopsy low power focus) Note periosteal new bone and subperiosteal and subcortical granulation tissue

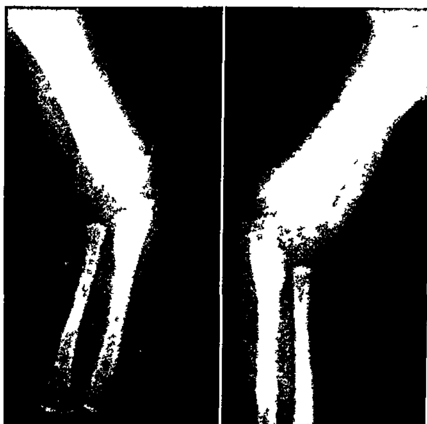


Fig 103 Syphilitic periostitis of humerus and forearm bones Case R R



Fig 104 (Left) Luetic periostitis of femur (acquired) Case L A (male) #45 3634 33 years November 1932 Pain in right thigh for six months fusiform enlargement of right femur X ray osteoperiostitis luetica Positive Wassermann Rapid recovery under antiluetic treatment a) November 1932 b) April 1934

Fig 105 (Right) Luetic osteomyelitis with pseudoparalysis Case R M (Courtesy Dept of Pediatrics State University of Iowa)

layers ensheath the entire bone, but usually they are the thickest at about the middle of the diaphysis and then taper toward the end (Fig 103)

The secondary or later type of periostitis appears as a reparative phenomenon of a healing process, particularly under antiluetic treatment This type fully resembles the syphilitic periostitis we see in the adult and acquired syphilis (Fig 104) The x ray picture differs in no way from that of a primary luetic periostitis It shows the same onion skin layers, the same network, the same trabecular formation, ending up in a thickened and hyperostotic cortex

The third feature of the early luetic bone lesion is the *diaphyseal osteomyelitis,* which is rather rare It occurs when the subchondral granulation tissue

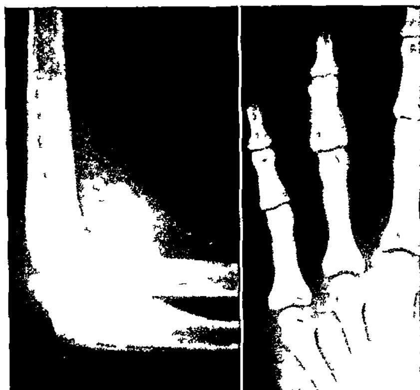


Fig 106 (Left) Tuberculosis of the elbow in case of congenital lues Case C H

Fig 107 (Right) Syphilitic dactylitis Note fusiform soft tissue swelling of the fourth digit

penetrates into the shaft The gummatous infiltration extends all along the bones and is most often seen in the radius and ulna The x ray picture shows areas of absorption, either diffuse or circumscribed, with typical punched out erosions (Fig 105)

That not all destructive lesions seen in a luetic are of syphilitic nature is demonstrated by the following case

Case C H (Female) #44 4504
Age 8 years Adm May 3, 1944

Entrance complaint of painful swollen right foot and right elbow of seven months' duration Patient was a known luetic There was the saddle nose and the Hutchin

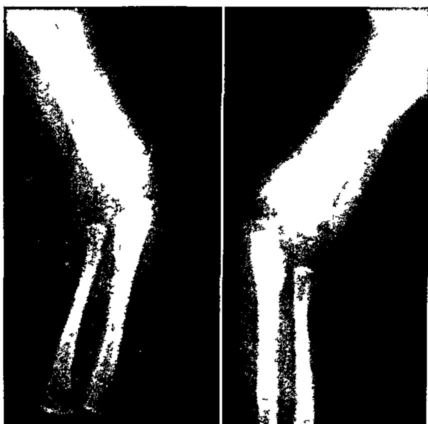


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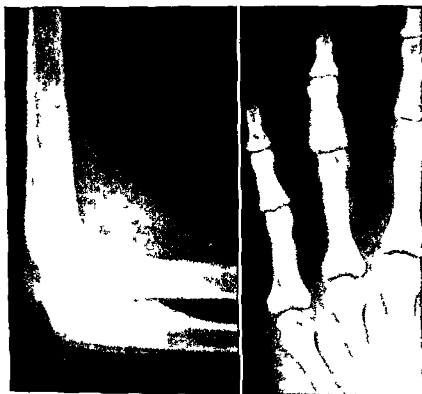


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son's teeth, the right foot was swollen, with an ulcer on the dorsum The right elbow was also swollen and immobile The x ray revealed destructive lesions of the right elbow and right astragaloscaphoid joint The condition of the right tarsus and the elbow was diagnosed as tuberculosis by biopsy and Guinea pig test She was treated by immobilization When last seen in June, 1948, the elbow was ankylosed at 120 degrees (Fig 106)

A special form of congenital diaphyseal osteomyelitis is the luetic dactylitis This is a diffuse, rarefying syphilitic osteomyelitis seen frequently in phalanges and metacarpals The diaphyseal osteomyelitis in congenital syphilis appears

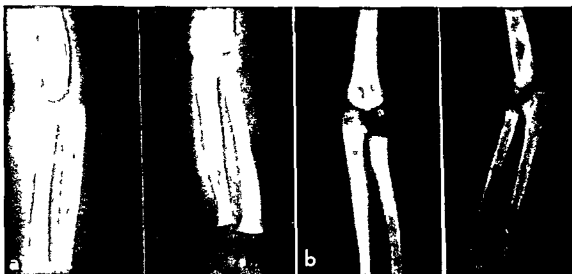


Fig 108 Osteochondriti and periostitis luetica with pseudoparalysis Ca e M K (female) #40 5973 eight months May 1940 Unable to use right arm for one month Luetic tigmata of nose and mouth hydrocephalus Wassermann Kahn and Klein positive X ray shows periosteal thickening of the humeri ulnae and radii Treatment with bismuth and ar phenamine prompt response a) Before treatment May 1940 b) After treatment November 1940 (Courtesy Dept of Pediatrics State University of Iowa)

later than the osteochondritis and periostitis The diaphysis is more consistently and more deeply affected than is the epiphysis The x ray picture shows first a decreased density in the small bones of the hands and feet, usually in the first and second phalanx The bones are not as dense as normal bone, and one notices a lattice arrangement of its internal structure (Fig 107) Sometimes there appear circumscribed cystic defects Combination of luetic periostitis with osteochondritis and pseudoparalysis is seen sometimes in infants (Fig 108)

2 The late hereditary syphilis of bone

In contrast to the fetal or prenatal syphilis, or that seen in early infancy, the favorite site of the late congenital syphilis is the shaft of the long bones It makes its appearance in childhood at the ages of five to fourteen years In the late congenital syphilis two essential pathological features are also noted, namely, the periostitis and the diaphyseal osteomyelitis The periostitis repre

sents extensive inflammatory bone production between periosteum and cortex, by the laying down of new periosteal bone in concentric layers The result is thickening of the cortex and narrowing of the bone and the marrow cavity

The diaphysitis or diaphyseal osteitis again involves more frequently the tibia In its initial stage there may be some softening of the bone, which results in a good deal of bowing before the thickening and condensation takes place

Syphilitic osteomyelitis of the skull affects mostly the frontal and parietal bones in localized or diffuse form Here, however, the phases of rarefaction are more pronounced, and syphilitic osteosclerosis occurs only later The first stages of rarefaction simulate Schuller Christian's disease The face bones undergo extensive destructive changes, such as destruction of the vomer, the saddle nose, dacryocystitis In fact, almost every bone of the head can be found involved The initial symptoms are dull, boring headaches not relieved by the usual remedies and often intensified by heat

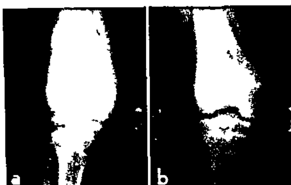


Fig 109 Late congenital syphilis gumma of femur Case M C a) Before treatment October 1933 b) After treatment April 1934

The gummas may appear as cortical lesions, the so called periosteal gummas, or they may be diffused, as in gummatous osteomyelitis (Fig 109) Necrosis and gumma formation may involve the subcutaneous tissues and skin, and may form sinuses with a cloudy, watery discharge These sinuses will persist indefinitely unless adequate treatment is administered

A history of recurrent attacks of pain and tenderness with intervals during which the patient is relatively free from symptoms is characteristic The pain is very often more intense at night, osteocopic, and is exaggerated by walking or exertion Objectively, one finds the enlargement of the bone, the deformation such as a saber tibia, and the luetic stigmata The most frequent of these are the saddle nose, the epitrochlear lymph nodes seen in 80 per cent, a positive spinal fluid found in 24 per cent, interstitial keratitis in 20 per cent, and Hutchinson's teeth in about 10 per cent

In typical syphilitic bone lesions, the diagnosis can be made easily from the clinical findings, the roentgenograms, and the serological tests However, there are atypical cases in which the problem is much more difficult, and a biopsy or a therapeutic test becomes necessary

The late congenital type of bone syphilis parallels in appearance the acquired form In comparison with the early type of congenital lues, it is comparatively rare, occurring probably in not more than 20 per cent of the cases of congenital lues

B THE SIGNS OF REPAIR IN EARLY AND LATE CONGENITAL SYPHILIS

1 Signs of repair in early syphilis

OSTEOCHONDRITIS AND EPIPHYSEAL SEPARATION

Spontaneous healing occurs, but under antiluetic treatment the repair is dramatic It is manifested by complete disappearance of x ray changes and by the undisturbed length growth of the bone The osteoblasts resume their former activity as they penetrate into the zone of calcified cartilage by means of the marrow tongues

HEALING OF THE SEPARATED EPIPHYSIS OCCURS IN TWO WAYS

A periosteal callus develops externally which causes a solid union with the diaphysis This may even occur while the separation through the granulation tissue is still in progress The callus is cartilaginous, it is thickest at the epiphyseal level, and tapers off toward the diaphysis Or, with the subsiding of the inflammation, a new process of proliferation and calcification of the cartilage begins The granulation tissue is then penetrated by marrow tongues from the bone, which carry osteoblasts Then the process of enchondral ossification occurs

HEALING OF THE PERIOSTITIS

Under antiluetic treatment, the different layers combine and consolidation occurs which, in the end, produces a thickened and hyperostotic cortex

2 Healing in the late congenital syphilis

The events are practically identical with what we see in acquired syphilis In the gummatous lesions the round cells diminish, fibroblasts increase, and cicatrization occurs Sometimes necrotic pieces of bone may be seen enclosed in scar tissue The gumma is surrounded by a wall of fibrous tissue with Langhans' giant cells situated between the fibers The fibrous scar may then be invaded by the marrow of the bone in the form of conical tongues, which proceed toward the center of the gumma The result is new formation of spongy fibrous bone within the gumma itself

II THE ACQUIRED SYPHILIS OF BONE

A GENERAL AND PATHOGENESIS

It is believed that the response to serological tests in adult acquired syphilis is variable As a rule, the percentage of positive tests diminishes with the age of the individual both in treated and untreated cases In view of this, it is of interest to compare the percentage of positive tests on patients with acquired syphilis who show osseous lesions Stokes, reporting a series of 239

cases of acquired bone syphilis, found a positive Wassermann reaction in 82 per cent In the experience of Speed, with over 100 cases of acquired bone syphilis, the Wassermann reaction was strongly positive in 90 per cent of the cases In cases where the clinical manifestations and the serological tests are at variance, the therapeutic test by antiluetic treatment almost invariably proves the reliability of the serological tests According to Stokes, among 63 cases which had received three or more courses of arsphenamine with mercurization, the Wassermann remained persistently positive in 50 per cent

Considering the relatively low incidence of neurosyphilis and cardiovascular syphilis, which should be recognized in the general examination of the patient, a persistently positive serum test is to a large degree associated with osseous reactions

The adult acquired type almost invariably results from sexual exposure The bone manifestations represent the tertiary stage of the disease They appear from one to two years after the initial infection In childhood, the acquired type results from other causes It may be acquired during parturition, nursing from a nipple with a venereal sore, kissing, etc As a rule, the acquired form which occurs in an otherwise healthy child has a much better prognosis than the congenital

B THE PATHOLOGY

In the tertiary stage the osseous lesion is typically osteoblastic and multiple Its most frequent sites are the tibia and ulna The saber shin which has already been described as a manifestation of late congenital lues differs in no way in the acquired type and is associated with the same periosteal bone apposition and endosteal proliferation The end result is that both periosteal and endosteal bone formation produce a partial or complete change of the normal cortical outline, as well as an obliteration of the medullary cavity

Late in the evolution of these hyperostotic lesions gummatous destructions occur They appear as sharply circumscribed areas along the cortex the entire thickness of which they may erode and so reach the medullary cavity (Fig 110)



Fig 110 Syphilitic periostitis and gumma formation (From H Beitzke *Handb d Spez Pathol Anat u Histolog* Berlin O Lubarsch and F Henke 9 477, 1939)

When we speak of syphilitic osteomyelitis,

we understand that the periosteum, the cortex, as well as the bone marrow are involved There is no essential difference between this osteomyelitis and the pyogenic type, except that the latter is much more accelerated compared with the chronicity of lues Here the sequence of events is usually as follows It commences with a chronic and inflammatory exudate composed of lymphocytes and plasma cells, which gradually organize, forming granulation tissue The fate of this tissue depends upon the healing capacity of the individual, and the efficacy and thoroughness of the antiluetic treatment If this treatment is adequate, complete healing ensues, and the granulation tissue becomes rapidly absorbed If there is no therapy and the patient has good general resistance, progressive fibrosis may occur with healing by scar tissue, but if the destructive effect of the Spirochaeta and its toxin predominates, necrosis of the granulation tissue occurs, with the characteristic pathological formation of the tertiary syphilis, namely the gumma The gumma may appear in the marrow, the cortex, the periosteum, or even the synovial membrane, and other joint components (Figs 111 and 112)



Fig 111 Syphilitic gumma in the marrow cavity (From H Beitzke *Handb d Spez Pathol Anat u Histolog* Berlin O Lubarsch and F Henke 9 495 Fig 24 1939)

Histologically, the gumma is an encapsulated formation of fibrous tissue In *the center is a firm elastic, yellowish mass, resembling hard cheese* The microscopic examination shows a periphery of dense fibrous tissue with numerous epithelioid and chronic inflammatory cells imbedded in it Compared with tubercles, giant cells are rare From the histological appearance alone, the differentiation between tubercle and gumma is not easy Massive destruction and sequestration are noted only in the skull This sequestration represents an acceleration of the destructive phase with subsequent rapid interruption of the vascular supply of a relatively large segment of bone



Fig 112 Syphilitic osteoperiostitis of skull (secondary lues) Note osteolytic areas in the skull (gummata)

C THE X-RAY FINDINGS

In the x ray the new bone formation appears in form of spicules laid down perpendicularly to the cortex and suggesting a lacework The best place to look for it is the anterior surface of the tibia, or the lateral aspect of the distal end of the ulna *Gummata appear* as characteristic punched out areas of rarefaction *They are always surrounded* by a halo of osteosclerosis Sometimes these translucent, punched out areas are extremely large and *few in number* Sometimes they extend way into the medullary cavity More often, however, they are small and numerous and are situated in the superficial cortex, giving it a rather stippled effect Since the *destruction proceeds slowly, sequestration* is rare, when it does occur, it almost invariably affects the cancellous bone Massive destruction of bone in syphilis is almost exclusively seen in the calvarium In the long bones, the periosteal reactions occur along the *entire course of the involved diaphysis* They eventually produce a very irregular, roughened cortical surface (Fig 113)



Fig 113 Healed syphilitic periostitis Case E L

D CLINICAL SYMPTOMS

Among 130 cases of bone syphilis Campbell found 98 acquired and 32 congenital It is believed that in 60 per cent of all cases of acquired syphilis, *bone and joint* symptoms appear sooner or later *In the syphilis* which is acquired in early infancy, the resulting lesions resemble more those of early congenital syphilis, but if the infection is acquired later in childhood, the changes are *similar to those seen in* the adult The typical bone lesions appear in the tertiary stage Their symptoms are usually dull, aching *pain poorly localized in the long bones* Later, one finds areas of localized bone tenderness, these may be transient, and they disappear *rapidly* under treatment

III THE CONGENITAL SYPHILIS OF JOINTS

A THE PATHOLOGY

1 Early congenital joint syphilis

Involvement of the joints in congenital syphilis is rare on the whole and occurs only in the later stages of osteochondritis There is no limitation of motion and no residual enlargement of the joints

2 Late congenital joint syphilis

As in the acquired form, congenital syphilitic arthritis of the later stages may again be divided into gummatous and nongummatous types The *non gummatous type* is found in the large joints of the knees, elbows, and shoulders and consists in diffuse swelling, similar to what we see in the secondary stage of the acquired syphilis The effusion is clear or moderately turbid, resembling that in nonspecific arthritis Rarely does one find Spirochaetae in the exudate The therapeutic test shows that salicylates prove ineffective, and antiluetic treatment gives a prompt response Under treatment, there is complete reces sion of symptoms

The second type of late congenital syphilitic arthritis is the *gummatous type* The gummata develop primarily from the synovial membrane, they develop slowly and are associated with effusion of a yellowish fluid The synovial membrane is hyperemic and is infiltrated with lymphocytes, especially in the perivascular regions Miliary nodes which start from the synovial membrane are seen in the villi Microscopically, these gummata represent jelly like grayish masses of granulation tissue, sometimes with irregular necrosis in the center

The joint cartilage remains intact for a long time, but in the course of de velopment some erosions appear which have a star-like shape They are in the middle of the cartilage, rather than at the margin When the case is treated, the defects fill out promptly with connective tissue A gummatous arthritis may develop from a gumma in the epiphysis, which extends into the joint Severe arthritic changes occur with the perforation of the gumma There is degenera tion of the cartilage and formation of a pannus from the synovial membrane which destroys the underlying cartilage, often leaving only small isolated islands The subsequent course of the gummatous arthritis is obliteration, partial healing or reconstruction If the obliteration is incomplete, small cavities of the joint may persist The joint constituents also may undergo considerable deformation which causes restriction of motion

It is also noteworthy that where the pannus is in contract with the old cartilage, a regeneration of the cartilage may be seen The new formed carti lage becomes sharply outlined against the old and uncalcified part When a recrudescence of the disease occurs, a new ingrowth of pannus may make its appearance This, again, can heal by metaplastic cartilage formation, so that the new formation will be separated from the old, preceding, cartilage layer

Sometimes several zones of calcification may be seen stacked one upon the other Thus a fair restoration of the articular surfaces may take place, and a satisfactory function of the joint may result, as long as the cartilage changes have not become too extensive

B SYPHILITIC BURSOPATHY OF VERNEUIL[6]

This is a painless enlargement of the knee which is due to inflammation of the extra articular bursae, sometimes seen in syphilitic joints The bursae are filled with fluid, but the joint surfaces themselves are not involved This condition follows a chronic course and is particularly resistant to treatment

C THE CLUTTON JOINT

A symmetrical serosynovitis was first described by Clutton It is essentially a hydrarthrosis of the knee joint which produces considerable distention and is usually observed in children between the ages of eight and 15 years It is commonly seen in congenital syphilis and is often associated with interstitial keratitis Robertson found this association in 80 per cent of the cases The joint appears enlarged, it is soft and painless, the patella is ballotting, and there is marked fluctuation One variety is the simple hydrops, and another, the combination with a proliferative growth of the synovial membrane The latter presents a reddish, swollen structure containing small gummata It is resistant to treatment

D THE CHARCOT JOINT IN CONGENITAL SYPHILIS[1]

We are accustomed to look upon the Charcot neuropathic joint as a sequela of acquired syphilis, occasionally, however, it is found in congenital cases That this neuropathic joint develops under traumatic influences from cerebro

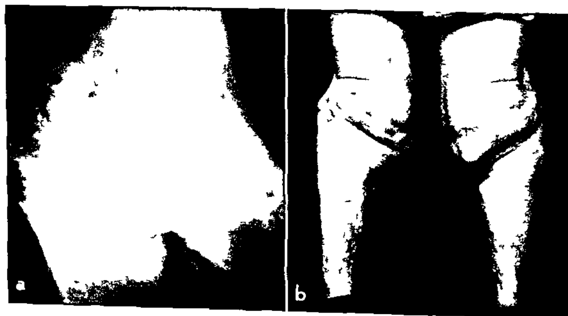


FIG 114a and b Bilateral Charcot s knee Case R S

spinal syphilitic lesions was the original theory of Charcot which has later been substantiated by experiments of Eloesser[3] The characteristic feature in the initial stage is atrophy, which leads to disintegration under the intervention of trauma The bone repair, however, is not maintained and the joint continues to become absorbed Disintegration proceeds and the joint finally melts away This destruction seems to be followed by an erratic attempt at reconstruction, a massive and disoriented production of bone, and the laying down of metaplastic bone in the soft structures The x ray shows fragments of bone in the joint, the muscles and the tendons The diagnosis is unmistakably established by the presence of abnormal mobility, infraction and collapse of the joint constituents, destruction of the articular cartilage sclerosis of the bone ends and the periarticular and periosteal bone formation It is, to all intents, identical with the Charcot joint which we see in acquired syphilis (Fig 114) (See Section B, Lecture V)

IV THE ACQUIRED SYPHILITIC ARTHRITIS

In combination with osseous lesions, acquired luetic arthritis is comparatively frequent, it occurs only rarely without them It has been stated that an adequately treated luetic never develops a true syphilitic arthritis The process usually begins insidiously as a synovitis The synovial membrane becomes thickened and the joint filled with fluid This is followed by the formation of villi Later, gummatous masses appear in the synovial membrane If the disease is not arrested then, the inflammation may go on to destruction of the articular cartilage Only rarely, however does any bone ankylosis develop

The transient changes which appear in the secondary stage of syphilis are different They respond readily to therapy and rarely if ever will produce any demonstrable radiographic findings or any permanent loss of joint function This arthralgia of the secondary stage is characterized by mild aches without joint effusion, and by the complete absence of any objective clinical or x ray findings Usually this synovitis is seen in the knee joint, but several joints may be involved simultaneously The clinical course is mild, and the condition responds readily to arsenical treatment

On the other hand the syphilitic arthritis which causes permanent changes belongs to the tertiary stage We may here distinguish again between a purely synovial and a gummatous form The simple synovitis takes a benign course The typical joint manifestation of the tertiary stage is the gummatous arthritis This is the type which results in considerable destruction and functional impairment It is usually due to the extension of a contiguous bony gumma which lies in the subchondral cancellous zone The destructive process may involve all the components of the articulation As it extends, it may cross the joint and involve even the opposite bone The syphilitic arthritis which arises from the synovial membrane itself produces extensive gummatous formations which finally destroy the synovial membrane the capsule and the articular surfaces, and ultimately result in disintegration of the joint

V THE THERAPEUTIC INDICATIONS

A CONGENITAL SYPHILIS OF THE BONES AND JOINTS

In young children it is somewhat difficult to apply intravenous therapy, therefore, other methods of medication must be adopted Sulfarsphenamine and neosalversan may be given safely intramuscularly in doses which vary with the age and weight of the infant, amounting to 1/10 to 1/15 of the adult dose The usual course consists in eight to twelve injections at intervals of four to seven days, followed by a rest period of three months During this rest period intramuscular injections of bismuth and mercury may be given

Furthermore, excellent results are reported by oral administration of stovarsol in cases of congenital syphilis This stovarsol contains 27 2 per cent arsenic, which is rapidly liberated in the acid medium of the gastric contents E C Mitchell of Campbell's Clinic reported on a series of 50 cases treated with stovarsol in which he obtained very favorable results, there were no untoward reactions The doses are 1/13 grain per kg body weight daily for the first week, 1/7 grain per kg for the second, 1/4 grain for the third, and 1/3 grain from the fourth week on The length of treatment is nine weeks to six months, with a rest period between treatments

Drug reactions are diarrhea and elevation of temperature, but these seem to clear up quickly, as do also the eczema and arsenical rash Sodium thiosulfate is the antidote in these cases

B ACQUIRED SYPHILIS OF THE BONES AND JOINTS

The same principles are applied here The simultaneous administration of mercury and arsphenamine is recommended by some, mercury being given by intramuscular injections of one of the soluble salts or by inunction The response of the syphilitic bone lesion to antiluetic treatment is prompt The symptoms disappear, and there is slow but steady improvement in the x ray appearance of the bone In infants, the epiphyseal lesions heal without causing any disturbance in growth or without producing any deformities In lesions of the shaft of the gummatous type, the normal architecture of the bone is restored completely

While congenital lesions of the joints are somewhat slow to respond to treatment, and this is particularly true of the so called Clutton's joint, in the acquired syphilitic arthritis the usual antisyphilitic treatment gets prompt and quick results

Surgical measures are seldom necessary in syphilitic bone lesions Should a secondary infection occur and a pyogenic osteomyelitis be superimposed upon a luetic infection, then the antisyphilitic treatment should be administered first, and the persistent bone infection should then be treated by whatever surgical measure is necessary

REFERENCES

1 Charcot J M *Arch de Physiol* *161* 1868
2 Clutton H H *Lancet* *1* 391 1886
3 Eloesser, L *Ann Surg*, *66* 201 1917
4 Klauder J V and Robertson, H F *J A M A* *103* 236, 1934
5 Parrot J M J *Maladies des Enfants* La Syphilis Hereditaire et le Rachitis Paris Masson & Co 1886
6 Verneuil A *Gaz Hebdom de Med et de Chir Paris*, *5* 609 1869
7 Williams J A *Lancet* *2* 977, 1882

Lecture IX

ON THE INFLAMMATORY DISEASES OF THE MUSCLES

IN THE study of muscle diseases, inflammatory or degenerative, one misses the solid pathological background which furnishes such a healthy foundation in other fields of clinical knowledge It is true that we have considerable information on the more terminal phases of inflammatory and degenerative changes of the muscle What is lacking is the knowledge of the intermediate and early pathology, the recognition of changes which are still reversible and in which the tissue reaction has not progressed beyond the possibility of repair

I ANATOMICAL OBSERVATIONS

A THE PASSIVE AND NON CONTRACTURAL STRUCTURES OF THE MUSCLES AND THEIR CIRCULATION

Nerve fibers and capillaries are carried by the interstitial connective tissue, the perimysium externum and internum, which forms a network in longitudinal direction The capillaries become very small and often are no wider than a single red blood cell, so that the circulatory inundation is very delicately balanced The normal vis a tergo is furnished by the muscle tone and by the rhythmic muscle contraction Consequently, in all conditions connected with a decrease of muscle tone, there is considerable nutritional disturbance in the parenchyma of the muscle Again, the larger vessels and nerves are carried in the bigger septa of the perimysium externum, these represent natural cleavages and are important in surgical technique They also are the principal sites of inflammatory cellular invasion The fibrosis which follows the inflammatory interseptal invasion produces constriction and ischemia of the muscle, and impedes its action both on the basis of circulatory deficiency and of purely mechanical interference with the free contractural play of the muscle [5]

The role of the perimysium internum is not entirely a passive one The collagen fibers surrounding the solitary muscle fiber are arranged crosswise, intersecting at acute angles, and they support the function of the fascia by controlling the form of the muscle in contraction Particularly in view of the fractional innervation of a muscle, this feature is of importance when only a portion of the muscle bundles is contracted at one time

The perimysium externum contains numerous elastic fibers which are arranged longitudinally and which assist the stretched muscle to regain its resting position Consequently, loss of elasticity of the perimysium always will accompany the parenchymatous degeneration and will contribute to the loss of muscle tension

B THE ACTIVE ELEMENTS OF THE MUSCLE ELASTICITY AND CONTRACTILITY OF THE MUSCLE FIBER

In inflammatory conditions the loss of elasticity proceeds rapidly with the interstitial scar formation, while contractility disappears more slowly, commensurate with the loss of parenchymal substance On the other hand, in degenerative diseases of the muscle we find that the contractile tone is lost early while the inert passive elasticity remains, the physical property of which has been discussed in the lecture on contractures

II PATHOPHYSIOLOGY

We must distinguish between muscle atrophy and muscle degeneration In atrophy there is a simple shrinkage with gradual disappearance of the muscle fibers, whereas degeneration is a parenchymatous reaction to toxic or inflammatory conditions The end stage of the atrophic and the degenerated muscle fiber is identical, because both finally succumb to complete absorption and disappearance, however, the intermediate phases show a vast difference in the course and particularly in the degree of regenerability

A THE SIMPLE ATROPHY

This again may be divided into neurogenic atrophy, due to injury or disease *of the spinal or peripheral motor nerve supply, and non neurogenic,* due to inactivity, arthrogenic causes, myogenic dystrophy, or inflammatory reactions The neurogenic muscular atrophy will be discussed with the paralytic disabilities as a feature of muscular paralysis The non neurogenic muscular atrophy may be divided according to its causation



FIG 115 Inactivity atrophy of muscle Giant cells and increase of sarcolemma nuclei (Muscle obtained from leg amputated for chronic osteomyelitis and soft tissue affection) Case H M

1 The inactivity atrophy

It develops slowly and is seen after immobilization or in amputated limbs (Fig 115) The atrophic muscle fiber becomes reduced in size but maintains its histological qualities It shows the greatest degree of regenerability, and recovery may take place even after years

The clinical corollary of this simple atrophy of disuse is shrinkage and flabbiness of the muscles, easy fatigability, and tenderness of the muscles on exertion The most damaged property of the muscle is the contractility, not the elasticity The weakened muscle corresponds to stimuli normally, except that its working efficiency is diminished in keeping with the decrease of its volume

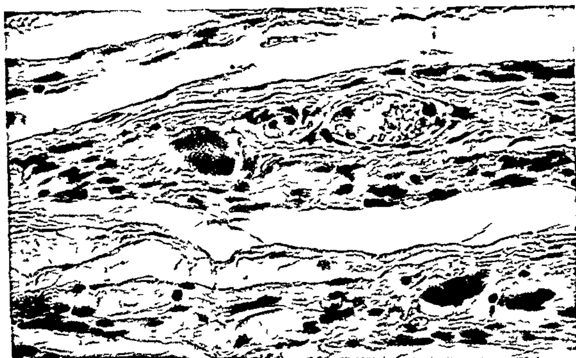


Fig 116 Simple muscle atrophy after tendon tear Muscle shows very pronounced atrophy cells are very thin but have somewhat preserved the transverse striation Sarcolemma cells remain and form typical muscle giant cells some fat tissue between the muscle bundles Case P W

2 The myogenic or tenogenic atrophy

It follows muscle trauma or tenotomy The atrophy of the muscles which follows tenotomy is particularly interesting There is a considerable weight loss of the muscle which in the first month may amount to as much is 40 per cent The contracted muscle is first hyperemic, then becomes anemic, and finally shows undulating fibers There is an increase of fat in the muscles, but not as much as we notice after nerve section (Fig 116)

3 The arthrogenic atrophy

It is a type of atrophy associated with inflammation or injury of the neighboring joint The traumatic reflex atrophy is best exemplified by the shrinkage

of the quadriceps following injuries or disabilities of the knee The inflammatory reflex atrophy progresses more rapidly and goes much farther than the purely traumatic atrophy, an instance of this is the atrophy of the thigh muscles in cases of tuberculosis of the knee

B THE TRUE MUSCLE DEGENERATION FROM INFLAMMATORY AND TOXIC CAUSES

Each of the different types of degeneration has its specific pathological features representing various degrees of degeneration and various prospects for regeneration

1 The cloudy swelling derives its name from the appearance of tiny albuminous particles in the muscle fiber The muscle loses its luster and the particles may cover up the striations The particles are situated in the sarcoplasm and not in the fibrillae

2 The granular degeneration is a more severe degree and may occur together with the cloudy swelling The difference is that here the albumin precipitate is larger The striations disappear, first the transverse, and then the longitudinal It occurs in inflammatory as well as in toxic conditions and already represents severe permanent damage In more advanced stages of degeneration vacuoles appear within the fibers, producing the so called hydropic degeneration *The nuclei of the sarcolemma remain intact* We find this degeneration in generalized diseases, in typhoid and tuberculosis, as well as in local disorders

3 The fatty degeneration In general conditions which cause cachexia, in severe degrees of phosphorus poisoning, in acute infectious diseases, in diphtheria, in inflammatory conditions of all types, in tumors, and after nerve section one observes fatty degeneration of the muscle which is different from the fat infiltration occurring in atrophic muscles The latter is a lipomatosis, that is, a deposit of fat between the muscle fibers in simple atrophy

4 The waxy degeneration or the degeneration of Zenker [4] The muscle appears pale, is reddish gray yellow, or even whitish in appearance, like fish meat, or sometimes turbid, like old wax Here, in contrast to the cloudy swelling, the contractile elements are involved In the course of time, rents appear in the muscle fibers which cause the fiber to crumble and to disappear altogether In these rents polynuclear leukocytes are found There is also a peculiar increase of the nuclei Finally, one sees tears in the sarcolemma through which the sarcoplasma escapes In toxic conditions such as typhoid, in phosphorus poisoning or in mechanical trauma, waxy degeneration may become general Local sites are the rectus abdominis, the psoas, the adductors, the shoulder, arm and calf muscles, and the diaphragm

5 The necrosis of muscles It develops principally from the shutting off of the blood supply, but can also occur in purely inflammatory conditions The necrotic muscle appears cloudy, pale and yellowish, and there is an absence of the nuclei

C THE RECOVERY FROM MUSCLE DEGENERATION

On the whole, the regenerability of the diseased muscle fibers depends essentially upon the condition of the sarcolemma If it is preserved, complete regeneration is possible The proliferation of nuclei is the first evidence of regeneration

Repair after waxy degeneration has already been studied by Zenker[4] (1863) As the fiber decays, there is a considerable increase of nuclei, and the whole sarcolemma appears filled with cells Those at the periphery become long, spindly sarcoblasts, and the round central cells disappear These sarcoblasts fuse with their dendrites and form long bands Gradually they show striations, at first longitudinal and then cross striations

III THE SPECIAL CLINICAL PATHOLOGY OF MUSCLE DISEASES

A THE CIRCULATORY DISTURBANCES OF THE MUSCLE

Complete ischemia is tolerated by the muscle for only two or three hours, after that, necrosis appears Pressure, for instance at the sacrum, may cause ischemia and necrosis of the muscles by compression of the superior gluteal artery with resulting embolic infarct

The most common example of muscle ischemia is the ischemic contracture of Volkmann In case of complete compression or severance of the cubital artery, massive necrosis occurs in a few hours If the venous backflow is impeded by the pressure of a subfascial hematoma at the forearm, degeneration and finally disappearance of the muscle fibers takes place more gradually

The passive hyperemia of the muscles of the lower extremities, which exists so frequently in cases of venous stasis, causes the muscle to assume a dusky color, and the blood vessels are engorged The muscle becomes edematous and the interstitial tissue proliferates and causes an apparent enlargement of the muscle, so called pseudohypertrophy Intramuscular hemorrhage, of course, is seen frequently in trauma, in hemophilia, or in embolism or thrombosis It may also appear in septic infections and in poisoning from arsenic and lead

B THE MUSCLE CHANGES DUE TO MECHANICAL CAUSES

Muscle Wounds

In muscle wounds, the edges separate and protrude the gaping is greater in transverse than in longitudinal wounds The severed muscle fibers retract in their sarcolemma Such an injury is usually followed by waxy degeneration and ends in partial necrosis of the muscle, including the sarcolemma and the perimysium In the subsequently developing scar tissue, one occasionally observes the deposition of lime salts

Regeneration Following Muscle Injury

Lesser cuts or divisions may regenerate by sprouting It starts from the

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nucleated sarcoplasm and consists in an amitotic proliferation of the nuclei Bud formations are observed on the sixth day after injury and may persist for six to eight weeks The striations of young fibrillae appear in the third week However larger muscle defects heal only by scars

In muscle ruptures, either total or partial the rupture occurs either at the musculotendinous junction or in the belly itself A muscle hematoma develops which becomes organized and forms nodes containing blood pigment and not infrequently calcium deposits Such a hematoma may be incompletely organized and become walled off, and the blood in the center may remain liquid for a considerable length of time A muscle hernia is a rupture of the muscle sheath causing protrusion of the muscle It is different from muscle rupture in that the enlargement becomes more noticeable on relaxation and lessens on contraction In muscle rupture, the opposite is true

The effect of gradual muscle stretching may be noted in the anterior abdominal wall in pregnant women There is a general distention or even a separation of the muscle fibers, and a process of degeneration sets in with loss of cross striation and crumbling of the contractile substance

C THE MYOSITIS OSSIFICANS

This is a bone metaplasia which may occur either in circumscribed or in more diffuse form *The diffuse form* is a rare pathological entity, and is of a progressive nature There is a gradual involvement of all the muscles and the underlying fascial and connective tissue, and in the most severe cases it may involve the entire body (Fig 117)

It starts with swelling of the muscle in childhood or adolescence and ends in progressive hardening and ossification The muscles first involved are usually the neck and shoulder muscles, then those of the back and chest As in the traumatic myositis ossificans, the bony masses are separated from the enveloping muscles by sheaths of firm connective tissue which penetrate into the bone lamellae of the mass The bone formation originates in the internal perimysium by metaplasia of a cellular and vascular tissue containing large, round and spindle-shaped cells In between the bone formations there are atrophic and degenerated muscle fibers It is noteworthy that the ossification involves not only the muscles but also the tendons and even the fatty tissue There is a decrease in the excretion of phosphates and calcium, together with a low blood calcium because most of the calcium is taken up by the ossifying muscles One also finds calcium in the kidneys and large intestines Extensive muscle calcification is also seen in paralytic conditions involving the cord, such as in transverse myelitis, and around Charcot's neuropathic joint

Calcification of the muscle often occurs after injury Contusions may produce necrosis of the muscle and calcium deposits in the necrotic tissue After trauma, particularly in the thigh muscles or around abscesses or in so called calcium gout one observes deposits of calcium in the muscle

The circumscribed form is usually of traumatic origin, from a blow, a cut,



Fig 117 Generalized myositis ossificans Case M W (female), #46 7163 23 years This patient was seen first in 1924 at age of 21 months when there was evidence of myositis ossificans of the upper dorsal and lumbosacral areas and scoliosis In 1938 had numerous hard masses in the region of the back and right hip In 1946 hard masses were noted in the left forearm and right biceps Histological examination showed fibrous tissue with metaplastic and enchondral bone formation a) Scoliosis b) Pelvis c) Forearm





a stab or a tear, or a sudden contraction of the muscle, or from repeated traumatism as, for instance, in the so called rider's bone of the adductors of the thigh (Fig 118) ›

The condition starts with an induration and painful swelling The first x ray sign appears after two or three weeks The bony mass is free in the muscle and is not attached to the periosteum, in contrast to an exostosis The tumor varies greatly in size and density It is round or egg shaped or flat, or it may appear in flakes or have a bizarre form, starting from several points

simultaneously It is encapsulated by firm connective tissue and is anchored by fibrous strands to the surrounding musculature Microscopically, one sees degeneration and atrophy together with new formations produced by budding myoblasts The calcium deposits are never in the muscle fiber itself (Gruber[9]) Bone formation occurs partly by metaplasia and partly by enchondral ossification The collagen fibers become wider and more homogenous, enclosing cells similar to those one sees in membranous bone formation On the other hand, one notices a transition of fibrous tissue to fibrocartilage, then to hyaline cartilage and finally to bone Osteoid seams surround the calcified bone



Fig 118 Myositis ossificans of the thigh



Fig 119 Localized myositis ossificans Case J W (male) #441463 20 years February 1944 This patient complained of shortening of the right heel cord for four years had hard mass of bone in the right gastrocnemius Slide shows muscle fibrous tissue and fibrocartilage with new bone formation

lamellae The bone marrow within the new formed bone is fibrous or fatty and contains giant cells, lacunae and osteoclasts, which explains the fact that very often these bone masses become absorbed (Fig 119) [3]

The treatment of myositis ossificans is rest and elevation of the limb, smaller pieces of bone may become absorbed spontaneously, for instance, those in the brachialis anticus of the elbow, or in the quadriceps above the knee Massage is contraindicated The orthopedic treatment should consist principally in immobilization Marottoli[13] advocates the use of plaster, which he considers the only prophylactic measure sufficient to avoid metaplastic ossification of the traumatized tissue [19]

So far as surgical intervention is concerned, it should be resorted to only in special cases in which conservative means have failed Above all things, it is necessary to wait until the new formed bone is entirely mature, and the period

of further bone formation has passed[18] Even when ossification has already taken place, the indication for immobilization still applies in order to accelerate the evolutionary cycle and to reduce the formation to a minimum Some authors (Marottoli[13]) advise x ray treatment at this stage in doses of 1200 to 1800 R[4] An unusual complication is malignant degeneration of the new formed bone tissue Peck and Brown,[14] reviewing the literature on myositis ossificans, found five cases in which the bone tissue underwent malignant neoplastic degeneration In addition, they cite three cases of their own

Our series of myositis ossificans of the elbow comprises 11 cases, the ages ranging from seven to 49 years Four cases followed fracture of the humeral condyles, two were preceded by dislocation of the elbow, one by fracture dislocation of the elbow, one by a Monteggia's fracture, and three by contusion of the elbow In all except one the symptoms appeared almost immediately after the accident Two cases were treated by physiotherapy, one with improvement Of three cases treated with braces or turnbuckle casts and physiotherapy, two improved, whereas the third remained stationary Three cases were treated operatively by removal of the bony mass, with definite relief obtained in two (Ponseti[16])

D THE INFLAMMATORY CHANGES IN THE MUSCLE

Compared with the great frequency of purely degenerative changes, true inflammatory changes of the muscles are rare

1 Suppurative myositis

It develops either directly by trauma or by contiguity from a neighboring focus of infection, and only exceptionally by the hematogenous route from a remote focus It is characterized by the usual accumulation of leukocytes by necrosis, the formation of abscesses and granulation tissue, and it finally heals with scar formation The muscle abscesses appear as doughy swellings containing a brown yellow pus which may or may not break through and produce a fistulous tract Multiple foci are often found in form of embolic abscesses and are characterized by granular degeneration of the muscles, by necrosis, invasion of leukocytes, and proliferation of the interstitial tissue (Fig 120)



Fig 120 Suppurative interstitial myositis Granulation tis ue is sub tituting the muscle cells which are atrophic and are separated from each other by markedly infiltrated fibrous tissue polys and lymphocytes are seen in the granulation tissue Ca e P O



Fig. 171 Dermatomyositis scleroderma Case L M (female) #43 14/0 50 years February 1943 Complaint of swelling in hands and feet four years ago lasting for three months brown discoloration of skin for three years and weakness of muscles Nodes on skin of elbows and knees extruding chalky material progressive stiffening of all parts of body Weight loss of 80 pounds dry adherent skin brown and scaly multiple nodules X ray absorption of distal ends of phalanges and fingers Diagnosis of calcinosis Treatment consisted in low calcium diet vitamins and iron resulted in considerable improvement with disappearance of nodes increased mobility and 80 pound weight gain a) Muscle atrophy of neck b) Stiffness and nodes of fingers

Gas infection is especially destructive to muscle tissue which readily succumbs to necrosis According to Aschoff,[1] gas gangrene develops only when the tissue is rendered anemic by pressure and thereby becomes susceptible to the infection, hence, the frequency of gas gangrene in the muscles of the buttocks, the shoulder girdle, and in muscle injuries due to shrapnel wounds It starts as an interstitial process which involves the muscle parenchyma secondarily (Hanser and Coenen[11]) The characteristic features are edema, gas formation, discoloration of the muscle to perhaps a dark blue or black, and finally the transformation of the muscle into jelly like mush Microscopically, the changes in the living muscle start with edema, fibrinous exudate,

and accumulation of leukocytes and gas bacilli This is followed by necrosis of the muscle as the bacilli enter into the sarcolemma No thrombi are found The muscle fiber finally breaks up, and the bacilli mass together in the interstitial tissue which separates the muscle fibrillae

The treatment of gas gangrene of the muscle is wide excision, debridement, and radical resection of the infected muscle tissue at the earliest possible moment In addition, massive doses of vaccine are administered, and extensive x ray radiation is carried out

2 Non-suppurative myositis

Nonsuppurative myositis consists of a painful swelling of certain groups of muscles and is associated often with edema of the skin, erythema, and urticaria Pathologically, it represents essentially a waxy or fatty degeneration of the muscles, with vacuoles in the fibers and inflammatory cells in the interstitial tissue Proliferation of the fibrillary nuclei and giant cells are seen The muscle appears pale, and shows punctate hemorrhages, it is also soft and fragile In the chronic stage, the interstitial proliferation prevails, crowding aside the muscle fibers

Similar edematous and inflammatory changes of the skin with additional endarteritis narrowing of the lumen of the small arteries, and proliferation of the intima are seen in *dermatomyositis* (Fig 121)

The so-called *periarteritis nodosa* is characterized by thickening and occlusion of the arteries, periarticular infiltration, and severe degenerative muscle changes (Figs 122 and 123)

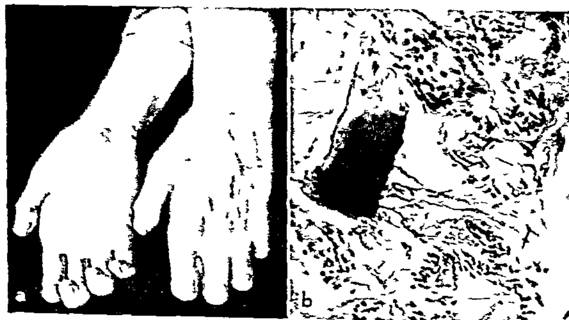


FIG. 122 Periarteritis nodosa. Case A. N (male) [illegible] 23 years, May 1947 Following acute febrile disease two years ago patient developed multiple joint lesions with redness and swelling diagnosis rheumatic fever Five weeks later complained of weakness of arms hands and legs paralysis of intrinsic muscles of hands. a) Clawhand deformity b) Slide from biopsy shows muscle degeneration, periarterial inflammation and thickening.



Fig 121 Dermatomyositis scleroderma Case L M (female) #43 1470 50 years February 1943 Complaint of swelling in hands and feet four years ago lasting for three months brown discoloration of skin for three years and weakness of muscles Nodes on skin of elbows and knees extruding chalky material progressive stiffening of all parts of body Weight loss of 80 pounds dry, adherent skin brown and scaly multiple nodules X ray absorption of distal ends of phalanges and fingers Diagnosis of calcinosis Treatment consisted in low calcium diet vitamins and iron resulted in considerable improvement with disappearance of nodes increased mobility and 80 pound weight gain a) Muscle atrophy of neck b) Stiffness and nodes of fingers

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of the muscle In this country, Rugh and Milch, as well as Plummer, Sanes and Smith,[15] described a case of hematogenous tuberculosis of the calf muscles More recently, Valls and Schajowicz [1] reported a case of muscle tuberculosis in a patient of 23 years, with a small pulmonary lesion

Even secondary tuberculosis is extremely rare, and it occurs then mostly by contiguity from a tuberculous bone or joint

Typical tubercles can be seen microscopically, developing between the muscle fibers and crowding them out while the muscle sheaths remain intact The nodes become confluent and large, showing caseation and liquefaction in the center The diagnosis is decided by the finding of the bacilli There is a sclerosing form in which there are streaks and strands of firm connective tissue containing numerous tubercles (Fig 125)

The disease begins with the appearance of a subcutaneous or relatively superficially situated tumor, which at first does not produce any functional disturbance The solitary tumor grows slowly, acquiring a round or fusiform shape, and is well circumscribed Its location in the muscle can be recognized by the fact that its mobility depends upon muscle contraction The differential diagnosis must include consideration of neoplastic formations such as the fibrolipoma and angioma, as well as trichinosis and cysticercus infection and such inflammatory conditions as syphilis and leprosy The treatment is surgical In most cases, the total removal of the tumor accomplishes cure

2 Leprosy A similar infiltration of the muscle is seen in leprosy Here the lesion appears in the form of wide strands containing round cell infiltration



Fig 124 (Left) Chronic interstitial myositis with fibrosis of the musculature and lymphocytic infiltration Case J T

Fig 125 (Right) Muscle tuberculosis (From Meyerburg *Handb d Spez Pathol Anat u Histolog* Berlin O Lubarsch and F Henke 9 394 Fig 34 1939)



Fig 123 Periarteritis nodosa Case J B (male) #47 6/04 63 years June 1947 Pain in joint and difficulty in walking for one year, flexion contracture of both knees a) Clawhand and intrinsic muscle atrophy left b) Slide from biopsy shows periarterial inflammatory infiltration

3 The muscle rheumatism or the so called myositis fibrosa

a Geloses

In this group belong the numerous geloses as well as the muscle scars described many years ago by Froriep[6] The essential feature is hyperemia and serous or fibrinous exudation in the muscle interstices These conditions are, as a rule, reversible, and it is therefore difficult to demonstrate them Graufhan[8] found small nodes simulating Aschoff's nodes in the interstitial tissue, and lymph follicles between muscle bundles Huselka[1] found foci in the muscles absolutely identical with Aschoff's bodies in cases of articular rheumatism and chorea The clinical manifestation is pressure tenderness and pain on motion of the muscles, usually accompanying or preceding joint pain

b Myositis Fibrosa

It is characterized by numerous connective tissue infiltrations, especially of the leg muscles, associated with pain stiffness and contracture White fibrous specks are found scattered through the muscles which are finally transformed into tough, scar like white tissue Microscopically, one finds proliferation of connective tissue and small accumulations of lymphocytes This is accompanied by the usual degenerative changes in the muscle fibers (Fig 124)

Fibrositis is the most common rheumatic disease It is most often of infectious or toxic nature Good[7] reported 125 cases, of which 40 8 per cent followed upper respiratory infection Draft and cold are the chief contributory factors Characteristic sites of these myalgias are the neck, shoulder, arm, back, lumbar region, hip, and leg They are often associated with, but not necessarily dependent upon, rheumatoid arthritis and osteoarthritis The usual treatment is heat, physiotherapy, massage, rest, and immobilization

E THE SPECIFIC INFLAMMATION OF THE MUSCLE

1 Primary tuberculosis of the muscle is hardly ever observed in man Hanke[10] found in the entire literature only 55 authentic cases of tuberculosis

The inner layer of the cyst is an amorphous substance surrounding the body of the parasite This condition must be distinguished from trichinosis, which gives smaller and more numerous shadows Phleboliths, although appearing perhaps in greater number, are usually round and are not likely to be confused with the shadows produced by cysticercus

2 The echinococcus There are up to now about 400 known cases of

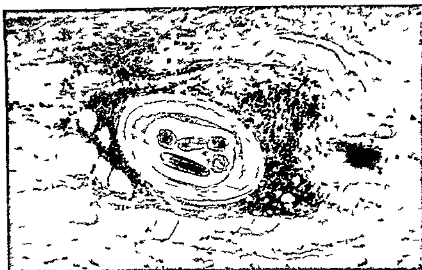


Fig 1?7 Trichina spiralis in muscle surrounded by some young fibrous tissue infiltrated by many lymphocytes

echinococcus in the muscle It more frequently involves the lower extremities The changes in the muscle are due to the pressure of the cyst, similar to those seen in the presence of a foreign body The cyst itself is surrounded by a wall of connective tissue the fibers of which penetrate between the adjacent muscle bundles The perimysium internum and externum of these muscles take part in the formation of this wall Under the pressure of the parasite these muscle fibers become atrophic and may entirely disappear When the cyst bursts, its contents penetrate the muscle and cause severe degenerative changes and violent inflammatory reaction, with the formation of abundant foreign body giant cells The parasite itself dies, and a scar is formed in the muscle which is similar to other muscle scars

3 The trichina is a special muscle parasite which enters through the gastric mucosa into the blood and the lymph stream It has a peculiar affinity for muscles, except the heart, most frequently involved are the diaphragm, intercostals, eye muscles, larynx, tongue, and a number of skeletal muscles It tries to locate near the tendinous insertion The young trichina, about 100 microns in length, grows rapidly and forms a spiral convolute The muscle itself develops reactive changes, first described by Virchow

A meralgia paraesthetica was described by M Bernhardt,[3] as being caused by trichinosis located in the muscles of the thigh The detailed anatomical changes (Askanazy) are as follows There is a peculiar degeneration of the muscle fibers which lose first the crosswise and then the longitudinal striations

and atrophic and broken up muscle fibers, between the fibrillae one finds the lepra bacillus The muscle fibers themselves show nuclear proliferation

3 Syphilitis myositis (Fig 126) This appears in form of a circumscribed gumma located in the muscle It is most frequently seen in the sternocleido mastoid, the thigh, biceps, triceps, masseter, and pectoral muscles Sometimes the gumma appears multiple and may attain considerable size It is a hard,



Fig 126 Muscle syphilis gumma (From Meyerburg *Handb d Spez Pathol Anat u Histolog Berlin* O Lubarsch and F Henke 9[1] 395 Fig 35 1939)

reddish white or yellow tumor and, microscopically, shows cell infiltration of lymphocytes and plasma cells, with thickening of the vessels due to prolifera tion of the intima which may even lead to complete occlusion Healing of the gumma occurs with a firm scar, sometimes even with calcification

F THE PARASITIC MUSCLE DISEASES

Of the many parasites which lodge in the muscles we shall mention only the most common

1 Cysticercus cellulosae, or Taenia solium, wanders through the stomach and the lymph and blood streams into the muscles and causes a violent reac tion of the muscle tissue Hemorrhage and discoloration of the muscle can be observed and is particularly frequent in the pectoralis muscle The initial symptoms are rarely recognized There is usually a long latent period before the cysts are recognized as subcutaneous nodes, or they may be accidentally found in the x ray picture when calcification has occurred This is seldom the case in less than five years

The radiographic appearance of the earlier stages suggests that calcium is first deposited in the residue of the fluid contents around the scolex The cyst itself undergoes considerable shrinkage during calcification The fully calcified cyst appears as an elongated, ovoid opacity in the long axis of the muscle

Lecture X

ON DUPUYTREN'S CONTRACTURE

I HISTORICAL

THE HISTORY of this deformity goes back to 1614 when, according to Coenen,[1] F Plater, an anatomist of Basle, described the condition as a dislocation of tendons To Baron Dupuytren[2] belongs the credit of having recognized the seat of the disease in the palmar aponeurosis It is interesting to follow Dupuytren in the detailed description of this condition which bears his name

In his dissection he noticed that the palmar aponeurosis was contracted and that from its lower portion there arose strands of cord which extended to the site of the diseased finger He observed that the superficial palmar aponeurosis formed by the expansion of the palmaris muscle and the prolongation of the anterior annular ligament of the carpus is extremely strong at its origin, but it grows thinner as it advances towards its lower edge It divides into four fibrous strips which are directed toward the distal ends of the four metacarpal bones There each of them bifurcates to allow the flexor tendons to pass, and each branch of this bifurcation is attached to the side of the phalanx These prolongations are more tense than the aponeurosis itself In his excellent monograph Coenen thoroughly supports the statement of Dupuytren that the seat of the contracture is the aponeurosis and that the latter sends various prolongations into the tendon as well as into the skin of the volar surface of the fingers

II THE ETIOLOGY

No definite fact regarding the cause of the disease has as yet been established Trauma or occupational strain in certain types of occupation may occasionally precipitate the deformity Dupuytren himself called attention to the fact that people of the most diverse walks of life are affected both heavy laborers and white collar workers Diabetes was long considered one of the causes, but it has been found to be more or less incidental There is likewise not sufficient evidence that infection is the cause of Dupuytren's contracture Some observers (Ledderhose[5]) emphasize the relation of the contracture to a concomitant arthritis deformans Both conditions have in common that they may be latent for a considerable length of time and are slow in development

In his series of 22 cases, 12 of which were bilateral, Ali Krogius[4] established a hereditary factor in not less than 4 cases, which led him to conclude that heredity plays a considerable role The idea of the deformity being a primary germ variation was advanced by Krogius It is based upon the observations on the behavior of the superficial volar hand muscles in mammals

and undergo fine granular degeneration This is followed by resorption of the muscle so that the trichina which was originally settled within the muscle fiber itself finally comes to lie in an empty sarcolemma sac (Fig 127)

After seven to eight weeks the encapsulation of the trichina is completed Granulation tissue surrounds the capsule, and this tissue may in turn take on fatty tissue, so that the parasite may finally become surrounded by fat The muscle in the surrounding areas appears reddish and firm The trichina, however, is not inert in this encapsulation, and may give rise to infection even after a number of years

REFERENCES

1 Aschoff L *Zentralbl f allg Path u path Anat* Supplement to Vol 27 1916
2 Askanazy, M *Aschoff's Path Anat Lehrb* Jena Ed 7 1 59 1928
3 Bernhardt, M *Zentralbl f d ges Neurol u Psychiat* Leipzig 14 242, 1895
4 Brailsford J B *Brit J Radiol* 14 79 1941
5 Braus H *Anatomie des Menschen* Berlin J Springer 1 69, 1921
6 Froriep R von *Die Rheumatische Schwiele* Weimar 1843
7 Good M *Ann Rheumatic Dis* 3 118 1942
8 Grauhan M *Anat Befunde bei Myositis Rheumatica* Inaugural Dissert Jena 1912
9 Gruber G B *Mitt a d Gren geb d Med u Chir* 27 76?, 1914
10 Hanke Hans *Deutsche Ztschr f Chir* 235 801 1932
11 Hanser R and Coenen H *Beitr z path Anat u z allg Path* 66 159 1920
12 Huselka T *Verhandl d deutsch Path Ges*, 17 470 1914
13 Marottoli O R *Bol Soc Arg Cir Ortop* 3 156 1943
14 Peck D C and Brown R R *J A M A* 19 776 1942
15 Plummer W W Sanes W W and Smith W S *J Bone & Joint Surg* 16 631 1934
16 Ponseti I *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa 16 D 23 1943
17 Recklinghausen F D von *Verhandl d deutsche Path Ges* 1 7 1899
18 Schede F *Munchen med Wchnschr* 76 508 1929
19 Smith N R *Brit J Surg* 26 780 1939
20 Thorndike A *A Bone & Joint Surg* 22 315 1940
21 Valls J E and Schajowicz F *Bol Soc Arg Cir Ortop* 3 1943
22 Virchow R *Virchows Arch f path Anat* 18 1860 32, 1865
23 Volcker F *Beitr z klin Chir* 33 1 1902
24 Zenker F A *Ueber die Veranderungen der Willkurlichen Muskeln in Typhus Abdominalis* Erlangen A E June 1863



FIG 1?9 Dupuytren s contracture Case M D

IV THE CLINICAL PATHOLOGY OF DUPUYTREN'S CONTRACTURE

The patient observes that it is more difficult to extend the fingers of the affected hand The ring finger soon contracts, and the contraction occurs first in the metacarpophalangeal, then in the midphalangeal joint The end phalangeal joint is never contracted, which is an argument against the tendogenetic idea which was formerly considered The ring finger is the one usually most severely contracted, the little finger, follows in order, and sometimes the middle finger The index finger is comparatively rarely involved

The ability to extend the fingers becomes more and more restricted In forcible extension of the finger the irregular strands become visible and palpable at the palmar aponeurosis As the contracture proceeds, wrinkles and furrows appear in the palm Under the skin one can feel numerous strands *running from the palmar aponeurosis toward the base of the finger* Dupuytren's contracture is prevailingly bilateral and is seen in more advanced age It hardly ever occurs before the third decade, and it is more prevalent in men than in women The development is slow and may extend over several decades

In our clinic 66 cases were seen between 1930 and 1946 (Koven[3]) The age range was 16 to 73 years, with an average age of 43 5 years Of these 83 per cent were males, and 17 per cent were females The presenting symptom was the appearance of nodules in the palmar fascia and their coalescence into bands, with gradual constriction and contracture of one or more fingers However, in some cases purely subjective symptoms preceded the objective symptoms, namely pain in four cases, tingling in two, and stiffness in three cases The initially involved finger was the fourth in 52 2 per cent of the cases, and the fifth in 38 2 per cent In general the deformity progressed slowly, and there were no reports of remission Frequently the progress was sudden and rapid and usually spontaneous in origin

In higher forms the fourth and fifth fingers show the development of special short muscles, but they are missing in the highest types from the anthropoid to man as well as in some lower forms of apes On the basis of these observations Krogius believes that the condition is one of developmental disturbance of the musculature with formation of a tendinous structure at the expense of an abnormal muscular "anlage"

The real shrinking process in Dupuytren's contracture is not in the fascial tissue as such, according to Krogius,[4] but it is newly formed tissue which undergoes shrinkage The cause of the disease is the unusual formation of regular tendinous strands within the layers of the palmar aponeurosis

III THE PATHOLOGY

The microscopic findings already mentioned by Coenen have been confirmed by Langhans and Jensen, and they are essentially as follows There is proliferation of the thick connective tissue without any inflammatory infiltration The cells show in general a direction parallel to the fascial bundle except in some places in which the arrangement of the fibers is more irregular (Fig 128) The blood vessels entering the strands are firmly surrounded by proliferating connective tissue These proliferations extend also into the walls of the vessel as well as into the nerves and tactile end organs The latter atrophy due partly to pressure



Fig 128 Dupuytren's contracture fibrous strands and arrangement of cells

The palmar aponeurosis is thickened, not by single isolated nodes, but by connected round strands running from the proximal portion of the aponeurosis to the contracted finger These strands are sometimes very firm, with a thickness up to 5 mm, and have a hard consistency (Fig 129) In the strands themselves it is possible to find cellular, almost tumor like accumulations of spindle cells as a typical new formation There are no signs of inflammation, nor any remnants of striated musculature

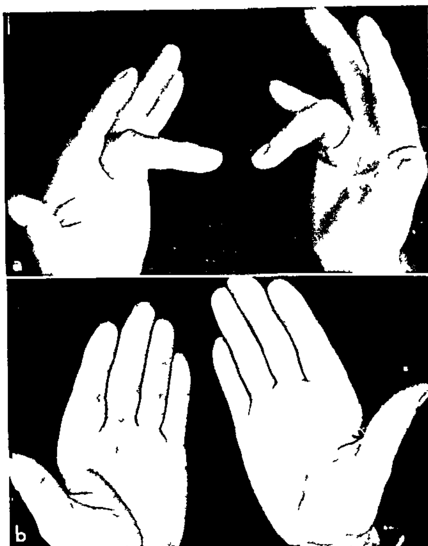


Fig 131 Dupuytren's contracture Case M W (male) #P 1127 53 years July 1936 Contracture of third finger right hand for six years and of fourth finger left hand for two year Resection of palmar fascia of the right hand November 1930 Resection of palmar fascia of the left hand was done in December, 1936 complete correction observation 11 months a) Before b) After operation

On resection one will find that the strands pass not only forward to both sides of the fingers, but also deep into the septa between the interossei All these strands have to be carefully resected Following the operation the hand is immobilized, and as soon as the wound is healed, careful stretching is carried out systematically by use of traction splints of the banjo type Following the resection of the palmar fascia and the straightening of the fingers, a gap often appears in the skin which cannot be closed by suture and which may require a skin flap plasty (Figs 130 and 131)

STATISTICS ON THE OPERATIONS (KOVEN[3])

Operative repairs were performed in 39 cases, of which three were combined with operation on the plantar fascia The operative approach was usually through the palmar creases and an L incision on the ulnar border of

V THE TREATMENT

A CONSERVATIVE

A new method of conservative treatment of Dupuytren's contracture is the use of vitamin E in form of tocopherols (Steinberg[7]) Steinberg attributes the effect of this drug to its ability to reduce the urinary creatine excretion He reported seven cases of Dupuytren's contracture of the hands with cure in six cases, the optimum dose was 300 mg vitamin E daily until the maximum improvement occurred He believes that vitamin E has a curative effect without surgery in early and moderately advanced cases, although in the more advanced and prolonged cases surgery might be combined with vitamin E to better advantage

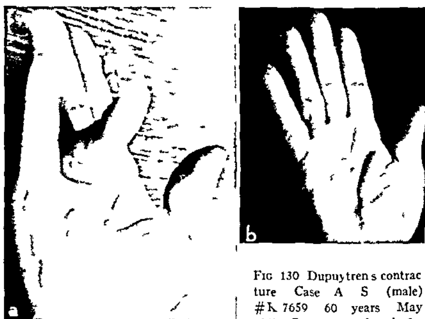


Fig 130 Dupuytren's contracture Case A S (male) #K 7659 60 years May 1947 Contracture fourth finger of right hand for 10 years Resection of palmar fascia was performed in July 1947 with excellent operative correction observation nine months a) Before b) After operation

B OPERATIVE

The early surgical attempts to correct the contracture were manifestly inadequate Dupuytren contented himself with subcutaneous tenotomies which he performed at different levels in the palm, in the metacarpophalangeal region as well as in the interphalangeal joints The subcutaneous tenotomies which had been gradually abandoned as inadequate were recently reinstated by J V Luck[6] in a more elaborate and apparently much more efficient method However, the usual procedure is the extensive resection of the palmar fascia from incisions running within the palmar creases To these an incision on the ulnar border of the hand is added so that a wide flap can be raised which exposes entirely the palmar fascia from the root of the hand to its expansions in the metacarpophalangeal region



FIG 134 Dupuytren's contracture of both hands and feet Case W B (male) #43-4?60 41 years March 1948 Patient complained of contracture of right hand for three years with painful nodules in palm for one year left hand contracture for two years *Typical* Dupuytren's contracture of hands bilateral Right foot shows thickening and tenderness of plantar fascia Resection of palmar fascia right in May 1943 and of palmar fascia left in March 1948 with capsulotomy of the proximal interphalangeal joint fourth finger a) Hands right postoperative left preoperative b) Soles



Fig 132 Dupuytren s contracture of palms and soles Case W L (male) #38 19097 38 years April 1937 Patient had nodules in palms for five years pain in palms for 34 years Resection of palmar fascia performed two years ago pathological report was subacute fasciitis Plantar fascia was also resected with good results Recurrence of palmar contracture Treatment of hands with x ray with good results in the left but both hands required reoperation good end result Gross pathology of plantar fascia revealed dense discrete nodule formation Micropathology revealed variable picture with many collagen fibers in ripples and whorls some sections cellular moderate vascularity with perivascular lymphocytic in filtration



Fig 133 Dupuytren s contracture of palmar and plantar fascia Case S D (female) #43 1605 46 years July 1936 Spontaneous onset of pain in feet for three months with tender nodules on both feet a) Contracture of third fourth and fifth digits for seven years Resection of palmar fascia on the left and bilateral plantar fasciectomy was performed in July 1936 Amputation of the right fifth toe for a painful neuroma was done in October 1936 Resection of palmar fascia on the right in November 1936 b) Gross pathology discrete nodular thickenings dense and white with longitudinal striations

Lecture XI

ON DISEASES OF TENDONS AND BURSAE

I ANATOMIC CONSIDERATIONS

CERTAIN anatomic features of the tendons and of the bursae surrounding them profoundly influence their reaction to traumatism and inflammation

a The tendons represent the continuation of the muscle, and are composed of tendon fibers separated by looser connective tissue The tensile force mediated through the muscle contracture is transmitted to the tendon Since the cross section of the tendon is so much smaller than that of the muscle, the ratio being in some instances 1 60, it is apparent that the unit strength of tendon tissues is taxed many times over that of the muscle

b The tendon itself is poor in blood vessels and nerves, except that it is endowed with proprioceptive end organs for the sensation of tension and position It makes little demand on the circulatory supply This, on the one hand, facilitates the survival in case of transplantation or transposition by operative means On the other hand, it is an unfavorable site for a primary infection, when infection occurs in the surroundings, the precarious circulation easily becomes shut off entirely and the tendon then succumbs to necrosis en masse, quite in contrast to the behavior in muscle tissue

c The vessels which enter into the tendon from the neighborhood are carried by the mesotendons Since the latter often are rudimentary or are missing altogether within the tendon sheath, it follows that the ability of the tendon to heal and to regenerate varies greatly with the location and the peculiar anatomical situation It is more favorable where the tendon has direct access to the blood supply of the surrounding areolar tissue than where it is isolated in a tendon sheath

d Bursae are established wherever friction is likely to arise Consequently, the bursae are eminently exposed to mechanical influences, and respond not only to actual injuries but also to occupational and other stresses

It is difficult to distinguish clinically between affections of the tendon sheaths and those of the neighboring bursae around the joints Besides, many bursae have direct communication with the joint itself, such as the subscapular bursa of the shoulder or the suprapatellar bursa of the knee Others are isolated, but are prone to perforate into the neighboring articulation in case of inflammation Still others again, such as the Baker's cyst of the knee, represent nothing more than a herniation of a synovial sac The clinical picture of the inflamed tendon sheaths and bursae varies so much with the location and the relation to adjoining structures that each situation requires its individual presentation

the palm Closure was usually effected, and when it was not possible, a full thickness graft was applied The results were as follows Excellent, that is, relatively normal anatomical and functional restoration, 12 cases or 38 per cent, satisfactory, that is, slight or very moderate pain and stiffness, and inability to fully extend or flex, 10 cases or 31 per cent

VI THE PLANTAR COROLLARY TO THE DUPUYTREN'S CONTRACTURE

In the literature only occasional mention was made of the fact that the plantar fascia may show similar changes The lesion was discovered in 1886 by Madelung Knavel, Koch and Mason have reported two cases since 1929 In a report by Dr J Vernon Luck[6] on Dupuytren's contracture no less than 14 cases of combined palmar and plantar lesions are discussed, the ages ranging from 24 to 72 years, with an average of 49 years Luck found in the tissue taken from the planta subacute or chronic inflammation, and in contrast to the the earlier observations of Ali Krogius, some foci of fascial necrosis were found These microscopic, multiple foci were in the process of being replaced by fibroblastic tissue growing in from the periphery on a scaffold of new capillaries Somewhat in contrast to the hands where strands are usually found, larger size nodules are more common in the feet, and the majority of lesions are located in the medial and distal portions of the plantar fascia (Fig 132)

Among our 66 cases of Dupuytren's contracture hands and feet were involved in 10 cases, and feet alone in one case Foot involvement was unilateral in six, bilateral in two, and unestablished in three cases

According to Luck,[6] the pain and tenderness is the presenting symptom, and it is episodic in character in some cases, being precipitated particularly by standing and walking The treatment is principally surgical, with resection of the plantar fascia similar to the resection of the palmar fascia in Dupuytren's contracture (Figs 133 and 134) Vitamin E therapy, the tocopherols, was used by Luck in two cases, but neither case showed any improvement in the contracture Nor was any improvement found from x ray therapy

REFERENCES

1 Coenen H *Ergebn d Chir u Orthop* 10 1170 1918
2 Dupuytren Baron *Leçons orale de Clinique Chirurgicale* Paris 1852
3 Koven L J *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *19 D* 25 1947 48
4 Krogius A *Acta chir Scandinav* 54 33 1922
5 Ledderhose G *Munchen Med Wchnschr* 44 254 1920
6 Luck J V Personal Communication 1949
7 Steinberg C L *M Clin North America* 30 221 1946

ish in color, and be infiltrated with pus, finally it succumbs to necrosis The extent of the purulent inflammation is then checked by a plastic exudate which attempts to seal off the infectious process, particularly at the narrow portion of the sheath The histological picture is that of a highly inflammatory condition, with leukocytes emigrating from the capillaries, thrombosis of the vessels, necrosis of the tendon, abscess formation and phlegmon The usual organism is the staphylococcus, more rarely the streptococcus

5) The rheumatic tenosynovitis Some types of articular rheumatism are associated with inflammation and effusion into the tendon sheath, the so called hygromatosis rheumatica Aschoff Gepel's bodies or nodes are found[1] Sometimes the infection is confined to the tendon sheath alone without the joints being involved, as in the syndrome of chronic rheumatic tenosynovitis[8]

b SPECIFIC INFLAMMATION

1) Tuberculosis Tuberculosis of the tendon sheath is the most common of the chronic inflammations of this structure Here we can distinguish three clinical variants (Garre[9]) One form is the serofibrinous type of Kauffmann, with formation of rice bodies Another is the fungus type, and a third is characterized by the formation of cold abscesses The purely serous form is rare The source of the rice bodies is the fibrin precipitation together with detached synovial villi Histologically one finds tubercles in the wall with fibrinous degeneration (Albertini) The fungus type shows more severe histological changes There is marked thickening of the wall of the sheath, which contains numerous tuberculous nodes One also sees accumulation of round cells and proliferation of the blood vessels The tendon itself finally becomes defibrillated, and granulation tissue invades its substance, penetrating into it by way of the mesotendon Finally the fungus masses liquify and form abscesses which soon perforate into the neighboring tissue as wandering abscesses or through the skin, establishing sinuses

2) Syphilitic tenosynovitis (Schuchart[4]) Verneuil was the first to mention syphilis of the tendons in his report on four cases Since then numerous reports have been published, especially by French observers An exhaustive study of this condition was made by Finger[6] The condition bears great similarity to tuberculosis The differentiation is made mainly on serological grounds There is a perivascular round cell infiltration, with small foci situated in the granulation tissue Large multinuclear giant cells similar to Langhans cells can be seen in the granulation tissue, but they are not characteristic for either syphilis or tuberculosis From the histological point of view, it is difficult to make the distinction The outstanding point seems to be the perivascular thickening and infiltration

3) Gonorrheal tenosynovitis[19] Next to the involvement of the *joints* it is the most frequent metastasis of gonorrheal infection, and it usually follows the joint involvement It is characterized by redness, *edema*, swelling,

II THE PATHOLOGY

A THE PATHOLOGY OF TENDONS AND TENDON SHEATHS

1 The inflammation of tendons and tendon sheaths

a NONSPECIFIC INFLAMMATION

Inflammation of the tendon itself is rare because it is poor in blood supply Usually the inflammation involves the sheath or the surrounding areolar tissue, which provides for the gliding ability of the tendon We distinguish the following types

1) The acute serous tenosynovitis, the so called rheumatic tenosynovitis, which follows trauma or is part of a generalized rheumatic infection

2) The acute fibrous tenonitis or synovitis crepitans, characterized by soft grating, so called "snowball crunching" Von Frisch[7] demonstrated that the seat of the disease is the surrounding connective tissue outside the sheath The changes consist in edematous swelling, hyperemia, and leukocyte and plasma cell infiltration The crepitation is explained by the friction of connective tissue fibrils in this edematous peritendinous tissue The most frequent site is the carpus, particularly the sheath of the abductor pollicis longus and the extensor brevis, also the extensor tendons of the foot and toes, and occasionally the peroneal tendon

3) In contrast, the so called *stenosing synovitis of de Quervain* [1] represents a thickening of the tendon sheath which interferes with its free play It is usually located at the abductor pollicis longus and the extensor brevis sheath of the thumb The snapping finger, or doigt a ressort, is another type of stenosing tenosynovitis, found mostly in the flexor tendons of the fingers It consists of a thickening of the tendon sheath in all layers The formation of fibrocartilage at the inner layer of the sheath has been observed in stenosing tenosynovitis[1] (Fig 135)

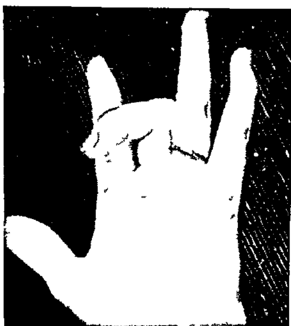


FIG 135 The snapping finger Case M C

4) The acute suppurative tenosynovitis Usually after a puncture wound, a synovitis of the tendon sheath develops, most frequently seen in the flexors of the wrist and fingers *Here is an example of the* rapid destruction of the tendon when the tendon sheath is filled with highly virulent infectious material In a few hours after the tendon sheath has become affected, the tendon will lose its shine become gray or green

herniation of the capsule, or they are synovial ganglia due to degeneration of the synovial sheath (Figs 137 and 138) At first they were considered simple herniations without degeneration, but it was Ledderhose[11] who established their true identity He showed that the ganglia are cystomas which develop through colloid mucous degeneration of the connective tissue The ganglia are lined by endothelial cells, but a strict division between this layer and deeper cells cannot always be made Older ganglia have definite similarity to the structure of the joint capsule, that is, they have distinct strata, an inner synovial and an outer fibrous layer Inflammatory processes may take place in these ganglia and cause enlargement and swelling They show perivascular round cell infiltration, an accumulation of spindle cells and sometimes giant cells

d Ossification of Tendons

New bone formation may occur in tendons, aside from the deposit of amorphous calcium, as described by Wrede [9] The bone formation occurs either at the point of insertion or at the musculotendinous junction, in the latter case it can be considered as a continuation of myositis ossificans, progressing into the tendon Occasionally ossification occurs within the tendon, without connection with either the insertion or the muscular junction There is no cartilaginous preformation The bone is derived directly from the connective tissue of the tendon, and is therefore fibrous bone Post-traumatic ossification occurs occasionally in the tendo achillis (Albertini) (Fig 139)



Fig 139 Compound fracture right os calcis with severance of Achilles posterior tibial flexor hallucis longus and flexor digitorum longus tendons and ossification of tendo achillis Case J K

Other frequent sites are the tendon of the triceps and the patellar tendon of the quadriceps The ossification of the tendon produces more or less difficulty in the movement of the joint, but rarely causes pain Although cases of tendon ossification after trauma have been reported repeatedly, it seems that trauma alone is not sufficient, a general disposition of the patient to bone formation is necessary

B THE PATHOLOGY OF THE BURSAE

1 The chronic inflammatory nonspecific hygroma

According to Albertini, the etiology of this hygroma is rather complex Older observers believed that it has an entirely inflammatory basis, but later observations seem to indicate that it is a process of liquefaction which de



Fig 136 Calcium deposit in the musculotendinous cuff in periarthritis of the shoulder Case S K

tenderness, and sometimes fluctuation The preferred localizations are the gastrocnemius, the extensor hallucis, the finger flexors, and the anterior and posterior tibialis tendons

The gonorrheal tenosynovitis may appear in serous, serofibrinous or purulent form The exudate contains diplococci There is marked hyperemia and round cell infiltration of the synovia, with loss of endothelium, followed later by the formation of massive adhesions The tendon itself degenerates and often ruptures (Melchior[17])

2 The degeneration of tendons and tendon sheaths

a Amyloid and Hyaline Degeneration

Amyloid degeneration is the great exception Only 1 case of this type was reported by Beneke,[1] who found it in connection with amyloidosis of the heart and of the large vessels

b Calcification

Calcification, on the other hand, is not unusual L Wrede [9] first described such calcification of the musculotendinous cuff in periarthritis of the shoulder joint (Fig 136)

c The Mucoid Degeneration

The mucoid degeneration of the tendon sheath is one of the principal causes of tendon sheath ganglia These ganglia are either arthrogenetic, that is,





Fig 138 Tendinous ganglion removed from extensor of the finger Case J W

←←←

Fig 137 Bilateral ganglion of the wrist Case W H

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velops on the basis of an arterial hyperemia, and the latter again is due to mechanical irritation This would lead to the formation of a fibrous mass which undergoes fibrinoid degeneration and then becomes liquefied It then comes to the formation of cavities characteristic for the hygroma (Albertini) Nevertheless, some of the changes we see in hygromas of the bursa are of inflammatory nature, as can be demonstrated by the thickening of the wall by inflammatory products For instance, in the case of a tuberculous rice body hygroma, we find numerous tubercles in the wall The hygroma, whether it is tuberculous or of nonspecific character, has an inner wall carrying villi and tabs The center of these villi consists of fibrous tissue, but toward the periphery one can see vascular granulations The surface of these villi or tongues is lined by endothelium Sometimes the tabs become separated from the wall and fall into the bursae as free rice bodies One may state, therefore, that the chronic hygroma is the result of a chronic inflammatory process, with certain additional degenerative changes within the thickened hygroma wall

2 **The chronic tuberculous hygroma**

The so-called rice body hygroma is a frequent occurrence in tuberculosis of the tendon sheath It is difficult sometimes to distinguish between the nonspecific and the specific tuberculous types, especially since the formation of rice bodies is not in itself a particular specific process and is found in conditions other than tuberculosis Housemaid's knee, for instance, is an example of a non tuberculous prepatellar hygroma

3 **The gonorrheal bursitis**

This is much more rare than gonorrhea of the joints or tendon sheaths Usually it occurs by extension into the bursa from a joint or a tendon sheath inflammation It is commonly the patellar and the subacromial bursae which are involved The pathological changes are similar to the inflammatory changes seen in the joint There is an acute stage with serous effusion and distention, and a chronic stage with thickening of the wall and proliferation of villi and tabs

4 **The chronic calcareous bursitis**

The Humeroscapular Periarthritis

Periarthritis of the humerus which follows injury to the musculotendinous cuff is only in part a traumatic disability, because in most cases there exists a certain predisposing factor of degenerative nature That inflammatory conditions occur in the subdeltoid bursa, and that they play a part in the so called periarthritis of the humerus has been known since the contributions of Duplay (1872)

We owe it to the investigations of Codman, Wrede, Brickner and others that this periarthritis is in most cases now recognized as a direct sequela of

injuries of the musculotendinous cuff Characteristically, these injuries are associated with deposition of lime salts into the tendon Inasmuch as these lime salt deposits often perforate the tendon and enter into the bursa itself, the calcareous bursitis should be looked upon as a sequela of a calcareous tendinitis of the musculotendinous cuff of the shoulder It has been shown that these deposits consist of calcium phosphates and carbonates, and constitute foreign bodies, they produce an irritation to which both the tendon and bursal sacs respond by inflammatory signs (Fig 140) The most plausible interpretations



Fig 140 Calcification of supraspinatus tendon and subacromial bursa Case J H

of these accumulations of lime salts is that they are secondary to necrosis of the tendon, just as in gout the accumulation of uric acid is secondary to necrosis of the cartilage

Calcification of ligaments (Fig 141) is not uncommon after injury, for instance, the calcification of the coracoclavicular ligament after coracoclavicular dislocation Another example is the calcification of the tibial collateral ligament of the knee joint, the so called Pellegrini Stieda's disease, which follows strain or injury of this ligament (Fig 142)



Fig 141 Calcification of coracoclavicular ligament following trauma



Fig 142 Pellegrini Stieda disease Case G B

velops on the basis of an arterial hyperemia, and the latter again is due to mechanical irritation This would lead to the formation of a fibrous mass which undergoes fibrinoid degeneration and then becomes liquefied It then comes to the formation of cavities characteristic for the hygroma (Albertini) Nevertheless, some of the changes we see in hygromas of the bursa are of inflammatory nature, as can be demonstrated by the thickening of the wall by inflammatory products For instance, in the case of a tuberculous rice body hygroma, we find numerous tubercles in the wall The hygroma, whether it is tuberculous or of nonspecific character, has an inner wall carrying villi and tabs The center of these villi consists of fibrous tissue, but toward the periphery one can see vascular granulations The surface of these villi or tongues is lined by endothelium Sometimes the tabs become separated from the wall and fall into the bursae as free rice bodies One may state, therefore, that the chronic hygroma is the result of a chronic inflammatory process, with certain additional degenerative changes within the thickened hygroma wall

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movement is generally limited by pain In tuberculosis of the tendon sheath the limitation of motion is more passive in nature Only as the infiltration of the sheath increases do we find that flexion contracture of the fingers becomes more marked, and that motion becomes more restricted

The simple nonspecific synovial cyst or ganglion which arises from the joint is sharply circumscribed and more often appears on the dorsal surface It is definitely painless and shows very little tendency to restriction of motion

TREATMENT

In children, tuberculous lesions often yield to simple conservative remedies, which should be the treatment of choice In individuals who already present other tuberculous lesions, especially pulmonary complications, it is best to proceed at first conservatively by immobilization, together with the injection of modifying fluids into the sheath Several cubic centimeters of iodoform glycerine or mentholated campor may be injected After injection, the member is enveloped in cotton bandage with a considerable amount of compression, and this bandage is retained for one or two weeks Although this is a long and tedious treatment, it still yields results in many cases, particularly in infants and children In all others, only long remissions are obtained, although the arrest may last for several years

The operative treatment is the rule for *adults,* and the method of choice is the complete extirpation of the diseased sheath This is done best under constriction with a blood pressure cuff, the sheath must be resected from one end to the other as completely as possible It is a difficult and laborious technical procedure, but it is an excellent operation if complete removal of the sheath can be obtained Otherwise, the results are anything but satisfactory, because recurrences are almost certain In cases in which there is involvement of the carpal bones, either preceding the tenosynovitis or secondary to it, resection of the carpus in combination with removal of the sheath must be carried out

Case J W (Male) #42 5952
Age 40 years Adm April, 1923

This patient came with a swelling of the left wrist which had existed for two years The examination showed an egg sized tumor of the wrist which was fluctuant The motion was free In April, 1923, the resection of the tendon sheath was performed But in July, 1924, there was a recurrence, and a large tuberculous tumor was removed from a deep flexor tendon In January 1926, another resection of the tuberculous tissue of the left wrist was performed The patient was relieved for six years until 1932, when there was evidence of carpal bone involvement In April 1932, a partial resection of the wrist was done, and this was repeated in May 1934, when a radical resection was performed The guinea pig test was positive for tuberculosis The pathological report showed tuberculous granulation tissue This patient was followed up until 1947, or 24 years after first admission He had no complaint The wrist was fused, and there was good functional use of the hand

III THE SPECIAL CLINICAL PATHOLOGY OF TENDON AND BURSAE DISEASE

A PATHOLOGY OF THE TENDONS

1 Inflammatory conditions

a Tuberculous Tenosynovitis of the Wrist

The existence of rice bodies in tendon sheaths was known to Dupuytren almost 100 years ago Its tuberculous nature, however, was not established until much later, with the finding of tubercles in the tendon sheath Both in its own right and as a complication of the osseous tuberculosis of the wrist the tuberculous tenosynovitis is of considerable clinical interest

Clinical Pathology

The onset, as a rule, is insidious and gradual Trauma, given as the cause in many cases, is only a contributing factor The synovitis appears as a tumor located at the volar surface, oblong in shape It has a firm consistency but is rather elastic The mass can be moved somewhat laterally, but in longitudinal direction it follows the contraction of the wrist and finger flexors If left to itself, the swelling increases, and the skin which at first is normal may become thinned out and discolored and even may ulcerate, thus establishing a sinus which emits a seropurulent fluid and occasionally a quantity of rice bodies At this stage the tendons are already immobilized in their sheaths, and a clawhand gradually develops

Spontaneous pain on the whole is not a prominent sign at the beginning of the infection in only two of our cases did we find it the presenting symptom But there are well localized pressure points which often indicate an underlying osseous lesion Limitation of motion is a rather late effect and on the whole it is remarkable how slowly it progresses In many cases a swelling exists for a number of years without a great deal of impediment to the movement of the fingers

Differential Diagnosis

One must distinguish between primary tenosynovitis and primary tuberculosis of the carpal bones In tuberculosis of the tendon sheath the swelling extends distinctly along the course of the tendon, that is, in the longitudinal axis of the limb While the motion is somewhat restricted, it is less so than would appear from the swelling The principal restriction is the passive extension of the fingers The pain is rather insignificant, and there is very little pressure pain, quite in contrast to the acute or pyogenic involvement of the tendon sheath

The site and form of the swelling differentiate it from tuberculosis of the carpal bones proper In the latter the primary swelling appears dorsally and is cylindrical so that the wrist is thickened in anteroposterior direction and

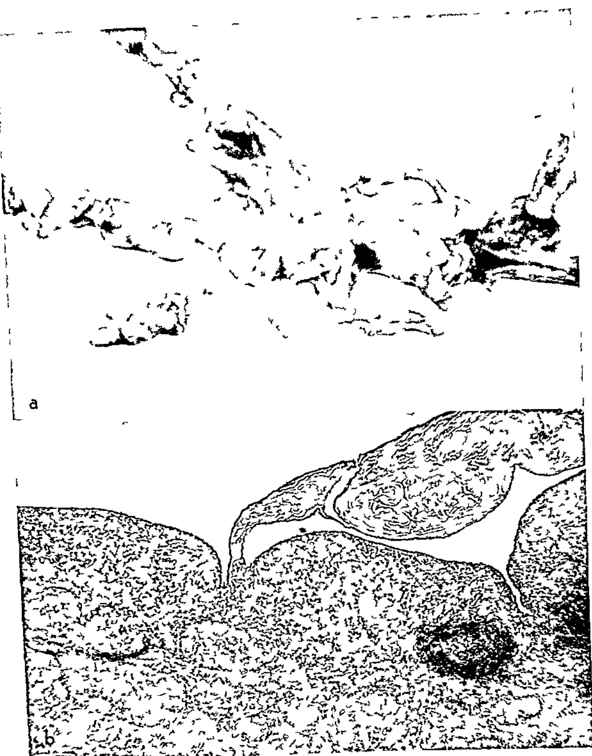


Fig 146 Tuberculous tenosynovitis of finger flexors Case L K (male) #K 10003 40 years February 1935 Soft swelling volar surface of right hand for three years swelling little and index fingers for two months Complete removal of the bursae of the deep and superficial finger flexors was performed in two sittings in February and March 1935 A third operation was performed in November 1935 for recurrent tenosynovitis of the finger flexors and removal of bursa of flexor carpi radialis a) Bursa after removal b) Slide showing rice bodies and tubercles



FIG 143 (Left) Tuberculous tenosynovitis of the digital sheath of the right index finger Case N I

FIG 144 (Right) Tuberculous tenosynovitis of the middle finger Case M W (female) #25125 54 years April 1932 Fusiform swelling over volar surface of right middle finger Biopsy showed tuberculosis Complete removal of the sheath was performed in February 1933 revealing a large number of rice bodies resulted in complete cure as seen in May 1947 or 15 years later



FIG 145 Tuberculous tenosynovitis of finger flexors Case N P (female) #44 5393 26 years December 1944 Healed tuberculosis of the lungs Pain in fingers and wrist and palmar swelling diagnosed as tuberculous tenosynovitis Complete removal of bursae of finger flexors flexor of thumb and flexor carpi radialis performed in February 1945 Pathological report confirmed tuberculous tenosynovitis No recurrence observation one year a) Exposed bursae b) Slide showing tubercles with giant and epithelioid cells



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In this case the tenosynovitis of the left wrist, which was undoubtedly tuberculosis from the beginning, was insufficiently resected, with the result that it extended into the carpal bones It was only after radical resection of the carpus that the disease was finally eradicated

In our series seven cases had radical resection of the sheath with ultimate good results, the observation time ranged from eight months to 12 years (Figs 143, 144, 145 and 146)

Conclusion Tuberculosis of the tendon sheaths of the fingers is a progressive disease which, in a large percentage of cases, ultimately ends with involvement of the carpus Early diagnosis of the condition is essential in the interest of timely intervention

With the exception of rare cases in children, or cases of multiple tuberculosis or pulmonary involvement the treatment of this condition should be radical and should consist in complete removal of the entire bursal sac In early tendon *sheath tuberculosis the outlook is not unfavorable if the treat*ment is radical However, incomplete removal of the tendon sheath is almost sure to lead to recurrences and possibly to ultimate involvement of the skeletal system We believe that very little can be expected in adults from injection of modifying fluids Even after operation, the tendency to recurrence must be considered for a long time, and the case must be subject to a long period of observation



Fig 147 Simple dorsal ganglion of the wrist Case K L C

b Nonspecific Ganglion of the Wrist

The ganglion of the wrist appears principally on the dorsal surface and is most frequently a round, soft and painless *swelling* which becomes more prominent when the wrist is flexed, and recedes on hyperextension Sometimes it can be displaced into the joint itself It interferes with flexion and extension of the fingers The extensor tendons separate and leave the ganglion between them, this mechanical disarrangement leads to early fatigue or pain on exertion The treatment is removal (Figs 147, 148)

c Stenosing Tenosynovitis

This is also known as Quervain's disease [1] *The condition is principally a* thickening of the tendon sheath and involves the long abductor and short extensor of the thumb The treatment is splitting of the tendon sheath or resection of the thickened portion

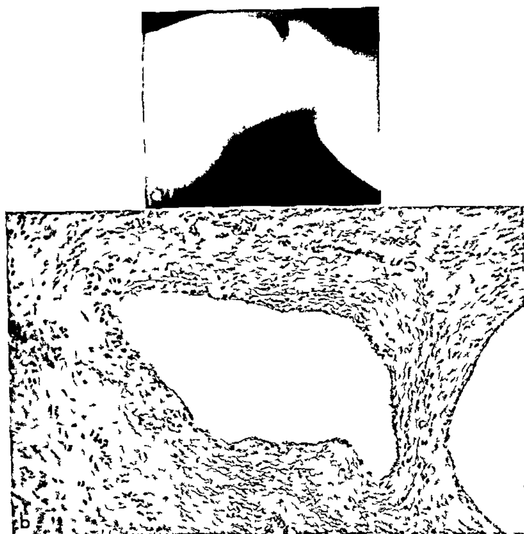


FIG 148 a) Simple ganglion of wrist b) Slide showing wall of ganglion Case J T

Case E O (Female) #57019
Age 41 years Adm September 17, 1945

In April, 1945, the patient noticed some pain in the right wrist In June she found that she could not use her wrist to write, and that she had some pain over the radial styloid region In the past three months the pain had increased All treatment, including splinting, bandaging, and physiotherapy failed The examination showed tenderness on palpation over the radial styloid, the abductor pollicis longus, and the extensor pollicis brevis tendons She was treated with a short arm cast with the thumb in abduction She remained in the cast for six weeks When seen in July, 1946, there was no pain in the hand, although some stiffness was present in the fingers In this case conservative treatment obtained improvement

Stenosing tenosynovitis has also been noticed in infants I Zadek [9] reports such cases of stenosing tenosynovitis of the metacarpophalangeal joint of the thumb, associated with pain on attempting extension Several cases of this kind have come under our observation There is no snapping on extension At operation one usually finds a constriction of the sheath of the tendon of the flexor pollicis

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Fig 147 Simple dorsal ganglion of the wrist Case K L C

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e NONSPECIFIC TENOSYNOVITIS OF THE LOWER EXTREMITY (Figs 149, 150)

The tibialis anticus tendon is a favorite location for the so called adhesive tenosynovitis In moving the tendon, one can feel the soft "snowball crunching" phenomenon and, on auscultation, a continuous and very soft grating sound is heard, like the rubbing of leather

Unless they are of long duration, these cases can be treated conservatively by immobilization with an elastic bandage and by physiotherapy

Case M N (Male) #60 114
Age 41 years Adm December 30, 1946

In August, 1946, this patient began to have swelling in the right knee, relieved by rest and aggravated by activity He also complained of pain in the right ankle on walking and standing The examination showed a swelling of the right knee with a patellar click Aspiration of the knee showed a clear fluid When he was seen again in July, 1947, the examination showed tenderness over the medial aspect of the right tarsal scaphoid, with a fluctuant swelling behind the medial malleolus In this case, the diagnosis of tenosynovitis of the right posterior tibialis was made An injection of 5 cc of 1 per cent novocaine into the tender area was made, giving complete relief The aspiration showed clear fluid from the sheath of the posterior tibial tendon This case was also treated conservatively with a leather anklet

Some results may be obtained in simple ganglia by purely conservative treatment, but the majority of cases must be operated on The operative results are uniformly good

In some quarters x ray radiation has been advised for the conservative treatment of ganglia F M Lyle[16] treated 21 cases all but two of which were located in the wrist The treatment was 1 5 erythema dose immediately over the tumor, one to three treatments being given He reports 17 good results in 21 cases In some of our cases, we obtained a subsidence of symptoms and a diminishing of the size of the tumor by simple rest and immobilization On the whole, however, we believe that ganglia of long standing are not affected by conservative treatment, and that they require operative removal

f SYPHILITIC TENOSYNOVITIS

This is a most exceptional condition, only a few cases of this kind are reported in the literature V L Schrager[3] reports a case of a man of 61 years who, for the past 20 years, complained of pain and periodic large swellings in the shoulder joint, wrist, and knee which were diagnosed as arthritis The condition was later recognized as a luetic synovitis because the Wassermann was 4 plus The patient was successfully treated by specific remedies This case developed a painless swelling in the upper portion of the left arm At operation a tumor about three inches in length was found in the tendon sheath of the long head of the biceps The pathological examination showed a perivascular lymphocytic infiltration with gummatous lesions The patient also had a similar condition in the *olecranon bursa*

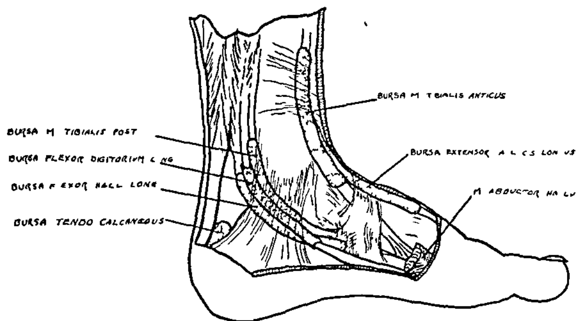


FIG 149 Tendon sheaths and bursae of the foot

longus or other tendons situated at the level of the metacarpophalangeal joint The treatment is surgical splitting of the narrowed sheath of the flexor tendon, and the results are uniformly good

d BICIPITAL SYNOVITIS OR TENOSYNOVITIS OF THE BICEPS TENDON

This is a common complication seen in periarthritis of the shoulder, and consists in involvement of the bicipital bursa It has lately come to be recog nized as one of the principal factors of the so called periarthritis



FIG 150 Chronic nonspecific tenosynovitis of the posterior tibial tendon sheath Patient had repeated swelling and pain about the posterior tibial tendon Diffuse infiltration of lympho cytes and plasma cells under the endothelial lining of the tendon sheath Case E R

The sheath of the biceps may be considered as an extension of the shoulder joint,[15] and an inflammatory condition involving this joint readily spreads into this sheath The inflammation is of the adhesive type and results in firm at tachment of the tendon mass to the bicipital groove It is only when this fusion has become solid enough to prevent any gliding of the tendon that the in flammatory signs subside An operative procedure becomes necessary if the case is of long standing, and if spontaneous fusion with subsidence of the symptoms does not occur The tendon should be released by operation, or it should be resected and reinserted into the lesser tuberosity

In the acute phases, inflammatory changes accompanied by pain can be recognized, they are undoubtedly due to the reaction of the tissue to the calcium deposits, as to foreign bodies A calcium deposit in the flexor carpi ulnaris was observed in 1 case (Fig 153)

Calcareous deposits in the tendo Achillis have been described repeatedly The same condition has been noted at the greater trochanter and in the tendons of the gluteals Kaplan[13] reports two cases of such calcareous tendinitis about the hip, one bilateral Here, also, the acute symptoms appear to be due to the acute inflammatory reaction about the deposit, with increased tension in the calcified area Release of tension usually relieves the symptoms, and the deposits can become absorbed Temporary relief can also be obtained by procaine infiltration of a 1 per cent solution

B PATHOLOGY OF THE BURSAE

1 The bursae about the knee joint (Fig 154)

a Anatomy

1) The prepatellar bursa lies between the skin and the patella As a rule, the interior of the bursal space is intersected by fibrous bands The bursa allows the skin to glide freely over the patella and to sustain pressure (Callander[3]) Because of its exposed position, this bursa becomes inflamed easily,

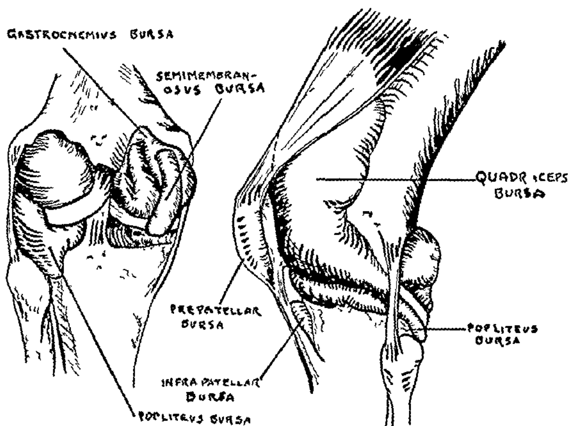


Fig 154 The bursae about the knee joint

2 Traumatic degenerative changes of tendons

a Calcium Deposits in the Subacromial Bursa

It is estimated that such calcium deposits are found in about 2 7 per cent of all persons (Bosworth), but only one third of these give a history of an occasional "hitch" or "twinge" in the shoulder at some time or other Small deposits of calcium may disappear *spontaneously, and the symptoms may subside* Large deposits always result in a painful shoulder sooner or later, even if they have remained quiescent for a number of years These large deposits do not disappear spontaneously, but usually require removal Codman has shown that these calcium deposits are primarily located underneath the floor of the bursa in the tendon of the musculo tendinous cuff, and that they cause bursitis only after they penetrate into the subdeltoid bursa The real cause of the deposit of lime salts is probably the primary injury of the tendons (Figs 151 and 152)



Fig 151 Calcium deposit in subacromial bursa

b Traumatic Calcareous Tendinitis in Other Locations

Sometimes a calcareous tendinitis is seen at the metacarpophalangeal joints Usually the condition is secondary to degenerative changes in the tendons, the adjacent ligaments, or the capsular structures The histological picture is that of degeneration of fibrous tissues The calcified areas are of variable size



Fig 152 (Left) Necrosis and calcification of supraspinatus tendon and impregnation with amorphous lime materia Case J H

Fig 153 (Right) Calcification of tendon of flexor carpi ulnaris Patient had pain in wrist for one year Tissue was removed from volar area of wrist where a calcium deposit had existed for one month Numerous small deposits of amorphous calcium are seen in areas of necrosis Case H D

In the acute phases, inflammatory changes accompanied by pain can be recognized, they are undoubtedly due to the reaction of the tissue to the calcium deposits, as to foreign bodies A calcium deposit in the flexor carpi ulnaris was observed in 1 case (Fig 153)

Calcareous deposits in the tendo Achillis have been described repeatedly The same condition has been noted at the greater trochanter and in the tendons of the gluteals Kaplan[13] reports two cases of such calcareous tendinitis about the hip, one bilateral Here, also, the acute symptoms appear to be due to the acute inflammatory reaction about the deposit, with increased tension in the calcified area Release of tension usually relieves the symptoms, and the deposits can become absorbed Temporary relief can also be obtained by procaine infiltration of a 1 per cent solution

B PATHOLOGY OF THE BURSAE

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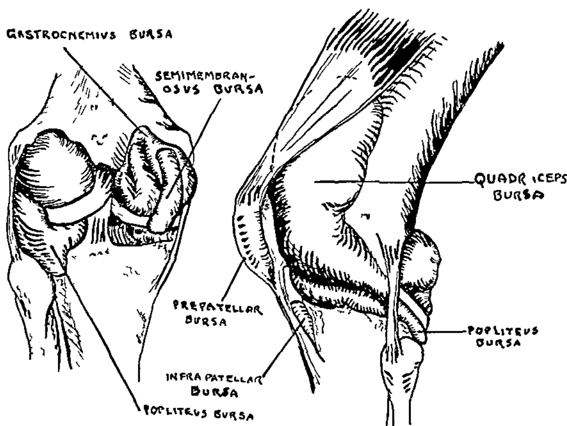


Fig 154 The bursa about the knee joint

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locking and no slipping, but there is a sense of insecurity That the mass lies outside the synovial membrane and under the tight fibers of the ligament can be demonstrated by careful examination

6) Bursae at the posterior aspect of the knee joint On the medial side there is a *semimembranosus bursa* between the medial head of the gastrocnemius and semimembranosus Besides, there is a bursa of the medial head of the gastrocnemius, between this muscle and the capsule Often there is a communication with the capsule

Case B E (Male) #37 710
Age 9 years Adm December, 1937

For the past four years the patient has complained of recurrent attacks of pain at the right knee During the painful episodes there was a noticeable swelling at the posterior aspect These attacks came every three months and lasted about two weeks They were severe enough to keep the patient awake The examination showed normal motion range in the knee, but there was a small painless tumor in the popliteal region which was lying lateral to the semimembranosus tendon The x ray was negative The patient was operated on, and a large cyst was found between the head of the soleus and the semimembranosus tendon, not communicating with the knee joint

7) Bursa anserina This bursa is located more distally under the conjoined tendon of the sartorius, gracilis, and semitendinosus, the so called pes anserinus (Fig 156)

8) On the lateral side there is a bursa situated between the biceps and gastrocnemius, the bicipital bursa, and also a small bursa over the lateral head of the gastrocnemius A small bursa is often situated between the tendons of the popliteus and the external collateral ligament, and another one at the lower angle of the popliteal muscle This latter often communicates with the knee joint



FIG 156 Pes anserinus bursitis of left knee Case E S

9) A special kind of pseudobursa is the extension of the synovial sac of the knee joint by herniation through the posterior capsule This appears most commonly lateral to the semimembranosus and gastrocnemius and is known as *Baker s cyst*

Case B A S (Female) #46 7143
Age 10 years Adm July 5, 1946

The patient s mother noticed swelling in back of the child s left knee 15 months before admission Twelve months before, a similar swelling appeared at the back

and effusion may terminate in suppuration A septic bursitis requires drainage by a wide incision In chronic cases the bursa becomes distended with a clear serous fluid presenting fluctuation This is the well known housemaid's knee, and it occurs from chronic irritation by prolonged kneeling This condition requires complete excision of the bursa, which can be done from a curved incision (Fig 155)

2) Pretibial bursa This is the bursa in front of the tibial tuberosity, between it and the patellar ligament A superficial pretibial bursa is sometimes located between the tendon and the skin

3) The suprapatellar bursa is really nothing but an extension of the knee joint cavity, as it is practically always in communication with the latter It takes part in all effusions of the knee joint Any expansion can be noticed readily at both sides of the quadriceps tendon

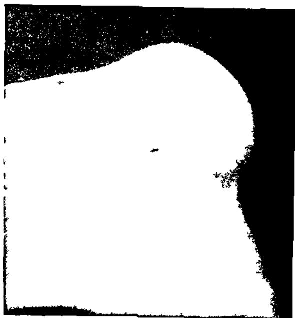


Fig 155 Prepatellar bursitis Case F J

4) At the medial aspect of the knee in the region of the tibial collateral ligament, a bursa is described minutely by Voshell and Brantigan [7] These investigators found bursae of variable number and size located deep to the tibial collateral ligament which did not communicate with any of the previously described bursae Sometimes they seem to be lying between the ligaments and capsule superior to the medial meniscus, sometimes, between the tibial collateral ligament and the medial meniscus, separating the meniscus from the parallel portion of the tibial collateral ligament, and sometimes, between the tibial collateral ligament and the tibia but in no way related to the meniscus [6, 27]

5) An analogous fibular collateral ligament bursa is described by I E Hendryson [10] The symptoms are a palpable tumor varying in size beneath the fibular collateral ligament near the middle of the parallel fibers This enlargement is always tender to pressure when the ligament is tightened by hyperextension The existence of this bursa under the fibular collateral ligament has been demonstrated already by Voshell and Brantigan,[26] and Anthony I de Palma[20] reports six cases The patient has the feeling of "tightness" within the knee joint, and the examination shows a small, tense round cystic mass on the anterior aspect of the fibular collateral ligament at the level of the knee joint The mass seems to dip beneath the anterior border of the ligament and then is seen again on pressure when the knee is flexed The treatment is complete removal It is significant for this condition that there is no

notices the position of the hip to be flexion, adduction, and external rotation There is a tilt of the pelvis which causes an apparent lengthening of the limb Hyperextension, abduction, and inward rotation produce pain Tenderness is present on pressure below Poupart's ligament and lateral to the femoral artery Iliopectineal bursitis may simulate hernia, hip joint disease, a perinephritic abscess, or a wandering abscess from tuberculous spondylitis Prolonged rest gives the best prognosis Incision is justified only in cases of suppuration

b SUBTROCHANTERIC BURSA

A bursa is situated between the greater trochanter and the tendon of the gluteus minimus One or two other bursae are situated between the tendon of the gluteus medius and the greater trochanter, and a third bursa is situated between the gluteus maximus and the iliotibial tract surrounding the greater trochanter This is an irregular, large multilocular space, the continuation of which covers the origin of the vastus lateralis There is also a bursa located between the gluteus maximus and the tuber ossis ischii, the so called bursa subcutanea ischiatica, which is not constant Another inconstant and smaller bursa is between the skin and the gluteus maximus tendon at the greater trochanter, the so called bursa trochanterica subcutanea



FIG 158 Subtrochanteric bursa complete removal Case C B

The signs of bursitis are usually local tenderness in the neighborhood of the greater trochanter, and a position of abduction and outward rotation Adduction and inward rotation cause pain The most accessible bursa is the superficial bursa trochanterica subcutanea Sometimes an acute subtrochanteric bursitis occurs, although this is rare Five such cases were reported by A Schein and O Lehman The patients complain of severe pain in the hip region, usually on weight bearing The typical finding is localized tenderness over the greater trochanter In some cases the x ray shows calcification lateral to and above the greater trochanter

The treatment in these cases consists in rest, cold applications, and novocaine injection, and in cases of chronic bursitis, complete removal of the bursa (Fig 158)

In a large proportion of cases the infection of the bursa is of tuberculous character Five such cases were reported by Donovan and Sosman [5] J D Wasserang [8] reported 18 cases of tuberculosis of the trochanter which involved the bursae The diagnosis can be made from the presence of pain, local tenderness, and the negative findings in the hip joint The aspirated material and

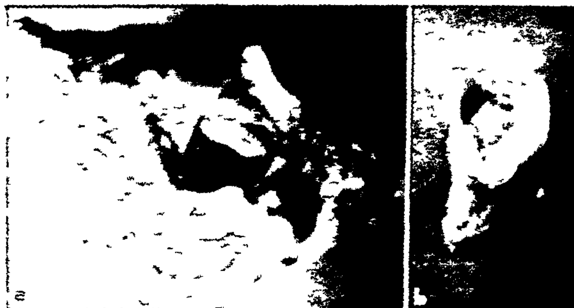


Fig. 157 a and b. Baker's cysts of both knees removed by operation. Case P. A. S.

[illegible] the right knee. The condition was painless. Examination of the right knee showed a small cystic tumor in the popliteal region. The left knee showed a slight increase in [illegible] and a similar mass 3 cm. in diameter in the popliteal region. The treatment was excision of the cysts on both knees (Fig. 157).

2. The bursae about the foot

There are numerous bursae about the foot. Aside from the tendon sheaths of the posterior tibial, the anterior tibial, and the peronei, there is a retrocalcaneal bursa behind the os calcis, between it and the tendo achillis; an [illegible] bursa lies underneath the os calcis, and a retro[illegible] bursa between the os calcis and the short flexor tendon. Furthermore, a small subcutaneous bursa is situated just on top and slightly behind the inner malleolus, and another bursa at the insertion of the tibialis anticus, between it and the scaphoid. Finally, a bursa is often found at the insertion of the peroneus brevis at the base of the fifth metatarsal, and occasionally one finds adventitious bursae.

3. The bursae about the hip joint

a. Iliopectineal Bursa

This bursa was first described by Fricke in 1834 and to date about [illegible] cases are reported. It lies between the psoas tendon and the iliofemoral and pubocapsular ligaments. Sometimes it communicates with the hip joint. This was the case in 14 per cent of all anatomical specimens examined by S. B. Chandler. The symptoms are pain in the hip region, often radiating in front of the thigh to the knee. The patient walks with a short stride, with knee and hip flexed, and the thigh is usually adducted and outward rotated. In later stages fullness and swelling appear in the groin. On examination one

22 SCHEIN, A and LEHMAN, O *Surgery* 9 771 1941
23 SCHRAGER V L *Arch Surg*, 48 423 1944
24 SCHUCHART K *Virchows Arch f path Anat* 135 394, 1894
25 VERNEUIL A *Gaz Hebdom de Med et de Chir Paris* (2nd Ser) 5 609, 1869
26 VOSHELL A and BRANTIGAN O C *J Bone & Joint Surg*, 25 121 1943
27 ——— *J Bone & Joint Surg* 26 793 1944
28 WASSERANG J D *J Bone & Joint Surg*, 22 1075 1940
29 WREDE L *Langenbeck's Arch* 99 259 1912
30 ZADEK I *J Bone & Joint Surg*, 24 326 1942

operative findings should give positive results In the series of 18 cases reported by Wasserang[20] all kinds of operations curettements excisions drainages and even sequestrectomies were performed

In our series reported by J E Milgram[1] there were 20 cases of subgluteal bursitis one-half of which were of tuberculous character The others were nonspecific or suppurative

Case W W (Male) #F-876
Age — years Adm February, 1931

The patient stated that since the age of twelve he had had a swelling over the right greater trochanter Eleven years before admission his left leg had been amputated for tuberculosis of the ankle The examination showed a discharging sinus over the right buttock The x-ray revealed a destructive process involving the greater trochanter The diagnosis was tuberculosis of the subgluteal bursa, following tuberculous osteitis of the greater trochanter In February, 1931 an excision of the bursa was performed After incision of the vaginal fascia and of some of the fibers of the gluteus maximus and medius an oval sac 2½ in long and 2 in wide was found. The sac was enclosed in dense fibrous tissue and filled with a [illegible] gray mass The inner wall of the sac was formed of gray granulation tissue An oval opening was found in the sac through which an instrument could be passed into a cavity in the femur In all particulars the sac corresponded to a subgluteal bursa. The bursal floor was excised en masse, together with the bursa and the sinus tract and a portion of the vaginal fascia immediately surrounding the sinus The wound was left open, and a vaseline drain applied. Pathological studies showed a tuberculous bursitis and guinea pig inoculation was positive. The patient was kept in a hip spica for four months at which time the wound had healed. When last seen one year later there were no complaints and he showed normal motion.

REFERENCES

1 Binkele R. [illegible] Beitr z Path Ges [illegible] 1929
2 Bosworth B M J.A.M.A. 116 [illegible] 1941
3 Callander C L Surgical Anatomy Philadelphia W B Saunders & Co 1936
4 Chandler S B Anat Rec [illegible] 1934
5 Dickson M S and Seidman M C Am J Roentgenol [illegible] 1912
6 Finger E A [illegible]
7 Frisch O von Arch f klin Chir 89 [illegible] 1909
8 Graff S [illegible] 1927
9 G[illegible] C [illegible] 1931
10 Henderson M S [illegible] J Bone & Joint Surg 2 [illegible] 1926
11 Hughes D C and Duncan F R Am J Surg 72 [illegible] 1946
12 Hutzler T [illegible] 1914
13 Kaplan L [illegible] 1941
14 Lindner [illegible] 1930
15 Lippmann R K Arch Surg [illegible]
16 Lyle F M J Bone & Joint Surg 23 1 2 1941
17 Melchior E D [illegible] 1919
18 Milgram J E J.A.M.A. 9 11 1932
19 Nicole G [illegible] 1912
20 de Palma Anthony F Am Surg 1[illegible]
21 de Quervain F [illegible]

22 SCHEIN A and LEHMAN O *Surgery*, *9* 771 1941
23 SCHRAGER V L *Arch Surg* *48* 423, 1944
24 SCHUCHART K *Virchows Arch f path Anat* *135* 394 1894
25 VERNEUIL A *Gaz Hebdom de Méd et de Chir Paris* (2nd Ser) *5* 609 1869
26 VOSHELL A and BRANTIGAN O C *J Bone & Joint Surg* *25* 121 1943
27 —— *J Bone & Joint Surg* *26* 703 1944
28 WASSERANG J D *J Bone & Joint Surg*, *22* 1075 1940
29 WREDE L *Langenbeck's Arch* *99* 259 1912
30 ZADEK I *J Bone & Joint Surg*, *24* 326 1942

Lecture XII

ON PRIMARY NEUROMUSCULAR DISEASES MUSCULAR DYSTROPHIES AND ATROPHIES

I THE PROGRESSIVE MUSCULAR DYSTROPHIES

MUSCULAR dystrophies are primary myopathies, they present a group of clinical entities the common features of which are onset at an early age, a hereditary tendency, a symmetrical involvement of certain muscles, and a propensity to progression There is very little known about the cause of this disease, although many factors such as trauma or endocrine disturbances have been considered The endocrine nature has been emphasized by Gibson, Martin and Buell,[3] on the grounds that the progressive pseudohypertrophic muscular dystrophy shows an occasional recovery at puberty when the glandular adjust

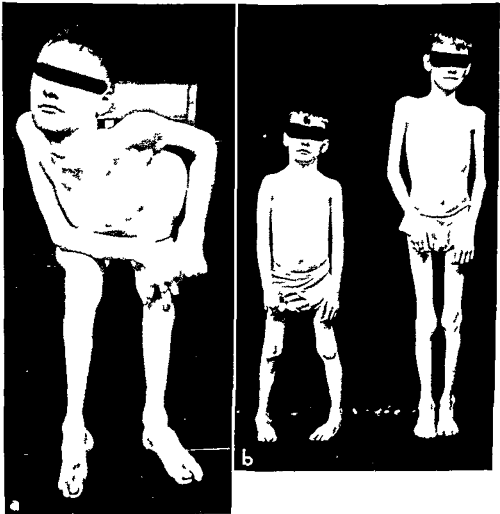


FIG 159 Progressive muscular dystrophy Erbs type a) Case R H *b) Cases H H and H H brothers of R H*

ments occur, and because improvement has been reported in some cases under endocrine therapy

The pathological picture is the atrophy of the muscles, the degree of which depends largely upon the stage of the disease We find the striations of the muscle fibers becoming indistinct, and the sarcolemma nuclei increased, with the progressing atrophy there is an increase of the interstitial tissue and a deposition of fat Later when the fat becomes absorbed, the muscular atrophy becomes more noticeable Changes are also noted in the blood vessels, the walls of which are thickened In the central nervous system, so far as the anterior horn is concerned, changes are secondary, in contrast to the situation in muscular atrophies

A THE SYMPTOMS

Usually the muscles of the trunk are involved first, and the extremities next The gait is unsteady and ataxic, and the patient becomes unable to rise to a standing position To do so he therefore adopts the peculiar expediency of climbing up on his thighs

Laboratory tests show an increase of creatine and a decrease of creatinine excretion in the urine due to the inability to burn up the creatine to its end products This lowered creatinine excretion was first noted by Spriggs,[5] who observed it in pseudohypertrophic muscular dystrophy as well as in myotonia congenitally and myasthenia gravis

Different types of primary myopathies or dystrophies are the following

1 The common juvenile or *Erb's type* has a familial tendency It starts in youth or in middle age and involves the shoulder girdle and upper arm, and is progressive The muscles show no reaction of degeneration, and there is an increase in the creatine output (Fig 159)

2 The *pseudohypertrophic type* of muscular dystrophy of *Duchenne*, also of unknown origin, occurs familially, it begins about the fifth or sixth year and is characterized by difficulty in the gait, by the peculiar rising mechanism already described and particularly by the very marked hypertrophy of the muscles of the calf (Fig 160 and 161) Biopsy shows the muscle fibers to be lacking transverse striation, the fibers are separated by young connective and adipose tissue There is hyaline degeneration of the sarcoplasm, and the

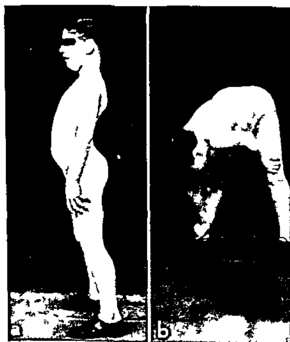


FIG 160 Progressive muscular dystrophy Duchenne's pseudohypertrophic type Case R B

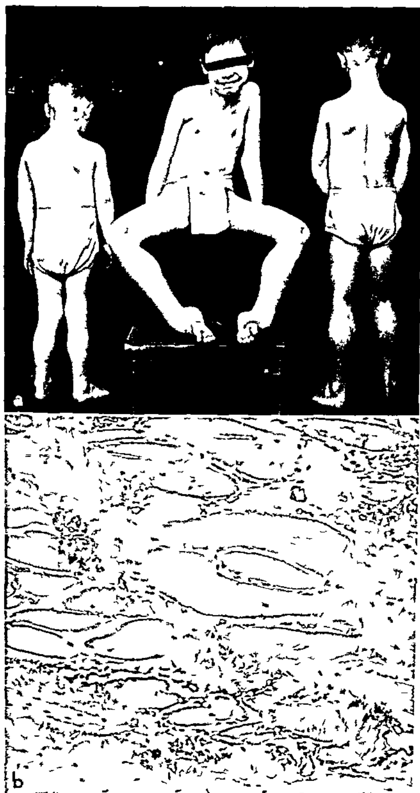


Fig 161 Progressive muscular dystrophy Duchenne's hypertrophic type a) Cases of brothers R W E W and R W b) Histological picture of muscle biopsy from right gastrocnemius Case R W (male) #38 1907? seven years Muscle fibers of different size with no transverse striation The muscle fibers are separated by young connective and adipose tissue sarcoplasm shows hyaline degeneration Sarcolemma cells are atrophic and scanty in number

sarcolemma cells are rather atrophic and scant in number

3 The *fascioscapulohumeral type of Landouzy Dejerine* This type also occurs familially and begins in infancy It involves particularly the fascial and shoulder girdle muscles It is progressive, but the muscles show no reaction of degeneration (Figs 162, 163, 164, 165, 166)

4 Other less common types are the *Leyden Moebius* type involving principally the pelvic muscles, and the distal type of *Towers Spiller*, in which the atrophy involves principally the muscles of hands and feet This condition is distinguished from the progressive muscular atrophy, because the muscles do not show any reaction of degeneration

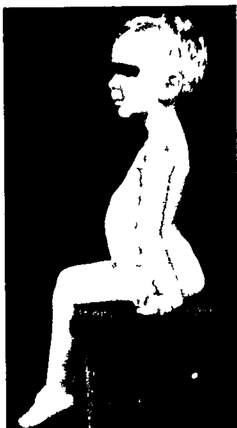


FIG 162 Progressive muscular dystrophy Landouzy Dejerine type Case R A

B THE TREATMENT OF PROGRESSIVE MUSCULAR DYSTROPHIES

The treatment of this condition has been eminently unsatisfactory so far There does not seem to be any way of halting the progress of the disease Two remedies which have been advocated rather lately are vitamins B and E Vitamin B in a synthetic compound was first advocated by Spies, Bean and Ashe[4] who noticed an increase in muscle strength in patients after administration of this vitamin Antopol and Schotland[1] reported some success in cases of pseudohypertrophic muscular dystrophy

Vitamin E is contained in wheat germ oil It was first suggested for progressive muscular dystrophy by H Blumberg[2] It is given in capsule form three times daily, each capsule containing three minims of the oil, for a total of nine minims per day, or the oil is administered in milk, and yeast tablets are added to the diet Cases treated successfully are reported by Stone[6]

II THE PROGRESSIVE MUSCULAR ATROPHIES

In contrast to the progressive muscular dystrophies, the atrophies are of central origin and involve the spine or the medulla oblongata The pathological changes involve particularly the anterior horn cells, with changes of degeneration similar to those seen in anterior poliomyelitis, that is, vacuolization, satellitosis, and neuronophagia The symptoms develop insidiously and slowly, involving particularly the small muscles of the hand The characteristic sign



Fig 163 Progressive muscular dystrophy Landouzy Dejerine type
Case J M

is the twitching, increasing weakness, and the disappearance of reflexes In contrast to the dystrophies, there is a reaction of degeneration Several types can be distinguished

A CLINICAL TYPES

1 The spinal muscular atrophy of *Duchenne-Aran* is a familial disease, and is due to degeneration *of the anterior horn and the anterior horn root* It progresses slowly, and characteristically involves the intrinsic muscles of the hand, particularly the opponens and the interossei It is a condition arising in infancy The involvement of the interossei and intrinsic muscles of the hand, leads to a symmetrical clawhand deformity There is reac tion of degeneration (Fig 167)



Fig 164 Progressive muscular dystrophy Landouzy Dejerine type Case R S

2 *Charcot Marie Tooth type* This is a type of progressive muscular atrophy with a familial and heredi tary tendency, and appears in the fourth or fifth year, it is character ized by weakness of the peroneals and quadriceps Its progress is slow, and it shows absence of reflexes The

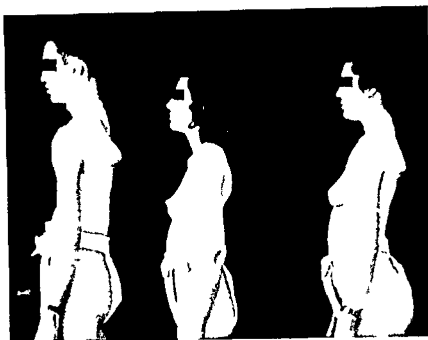


Fig 165 Progressive muscular dystrophy Landouzy Dejerine type Family R S C S and V S

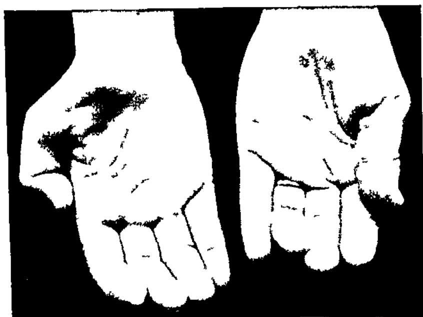


Fig 166 Progressive muscular dystrophy Landouzy Dejerine type Also thenar atrophy Case V S

pathology is degeneration of the Goll's tract, and peripheral nerves, with fatty degeneration of the muscles (Fig 168)

3 The familial progressive muscular atrophy of *Werdnig-Hoffman* is a type with familial occurrence, and appears in the first year, it involves particularly the trunk muscles It progresses rapidly and usually ends in death There is tremor and reaction of degeneration The pathological basis is degeneration of the anterior horn

4 The *Friedreich's ataxia* was first described by Friedreich, in 1863 This is a strongly hereditary form which occurs usually in juvenile ages, from 12 to 15 years, it is characterized by progression and ataxia There is a positive Babinski, and the tendon reflexes are diminished The characteristic signs are the cavus deformity, involvement of the speech, and nystagmus The pathological basis is degeneration of the posterior column, the posterior roots, the ganglia, and the column of Clarke, and degeneration of the muscle fibers

5 The *congenital myotonia of Thompson* is a strongly hereditary disease



FIG 167 Progressive muscular atrophy, Duchenne Aran type Atrophy of thenar and intrinsic muscles Case D B (female), #A 6577 2 years July 19?1 At the age of nine months patient was unable to hold up her head and was extremely undernourished at one and one half years her legs were still weak and underdeveloped She had hyperextended knees and flabby musculature atrophy of the trunk and shrinkage of muscles of the hand *There was weakness of the seventh cranial nerve with inability to* raise the head and weakness of the arms and legs

appearing in early childhood There is no degeneration of the muscle fibers The latter show a strong rigidity and involuntary inhibition of motion The faradic and galvanic irritability is increased The condition has the aspect of spastic paralysis, with strongly increased muscle tone

6 The *amyotonia of Oppenheim* is, in contrast to the congenital myotonia of Thompson, a disease characterized by flaccid arms and legs, and particularly atrophy of the intrinsic muscles It is acute and progressive The electric irritability is definitely decreased, as are the reflexes The pathological basis is atrophy of the anterior horn associated with atrophy of the peripheral muscles

Several types of spinal cord lesions must be distinguished from progressive muscular atrophy There is first the *chronic anterior poliomyelitis* which develops more rapidly, and in which paresis antedates the muscle wasting

The *amytrophic lateral sclerosis* often considered a variant of progressive

muscular atrophy, is characterized by the same progressive muscular atrophy, but in addition there are signs of lateral tract lesions in the form of spasticity, increased reflexes, and positive Hoffmann and Babinsky signs *Syringomyelia* may also produce similar findings, except that it also shows a characteristic disturbance in sensation, particularly in the thermic sense

B TREATMENT

As in the muscular dystrophies, no satisfactory method of treating this disease has been found The medicinal treatment is entirely tentative Strych



FIG 168 Progressive muscular atrophy Charcot Marie Tooth Type Case M F (female) #39 15315 8 years December 1947 At age of seven years patient had foot drop a steppage gait and atrophy of both calves The peroneals and anterior tibialis showed no response to *electrical stimulation faradic or galvanic* no sensory changes Condition was slowly progressive hands not involved Neurology concurs in diagnosis Charcot Marie Tooth peroneal paralysis One sibling has similar leg difficulty but has not been examined here

nine is used in rather large doses, of 1/30 gm daily Tonics such as elixir of iron, quinine, and strychnine, *in doses of 4 cc, or in the form of ferrous solu*tion given five to six drops three times a day, have been applied Their effect is doubtful Ample rest, avoidance of overexertion and fatigue, and proper hygienic measures, including a high caloric diet restricted in carbohydrates, seem to have some effect in slowing down the progress of the disease

REFERENCES

1 ANTOPOL W and SCHOTLAND C E *J A M A* *114* 1058 1914
2 BLUMBERG HAROLD *J Biolog Chem* *108* 227 1935
3 GIBSON R B MARTIN F G and BUELL M V R *Arch Int Med* *29* 82 1922
4 SPIES T D BEAN W B and ASHE W F *J A M A* *112* 2414 1939
5 SPRIGGS G I *Biochem Ztschr* *2* 206 1907
6 STONE, SIMON *J A M A* *114* 2187 1940

Section B

DEFICIENCY AND DEGENERATIVE DISEASES OF THE LOCOMOTOR SYSTEM

Section B

DEFICIENCY AND DEGENERATIVE DISEASES OF THE LOCOMOTOR SYSTEM

Lecture I

ON AVITAMINOSIS

I VITAMIN D DEFICIENCY

THE principal effect of vitamin D deficiency upon the skeletal system is that the absorption of calcium which depends on the vitamin D becomes impaired Bone diseases caused by insufficient intake or absorption of vitamin D are rickets, infantile or late, osteomalacia, and senile osteoporosis The latter will be discussed with the senile diseases of bone

A RICKETS

Animal fats and especially cod liver oil have been found to be a rich source of vitamin D It is likewise known that the ultraviolet rays of the sun transform the cholesterol of the body fat into a substance called ergosterol, which is capable of causing vitamin D to be absorbed into the body The normal sources of calcium are many vegetable foods and notably milk and milk products

The essential role calcium plays in the metabolism of bone needs hardly to be emphasized Bone contains 98 per cent of the calcium of the body and 65 per cent of the body phosphorus (Arazoza[1]) The calcium in excess of what is absorbed is eliminated in the feces, and a smaller qualtity in the urine The tendency of the blood to maintain its calcium level causes it to draw on the calcium storage of the bones whenever the blood level becomes low due to insufficient absorption

The calcium phosphates are relatively insoluble in alkaline, but soluble in acid solution, this solubility necessary for their absorption is mediated through vitamin D Since this vitamin is soluble in fat, which is the vehicle carrying it, it follows that the function of the pancreatic and hepatic secretions is necessary for absorption of the fat containing the vitamin Thus, a lack of calcium in the bone may be due to a lack of ingested calcium, or of vitamin D, or to the inability of the intestines to absorb fat because of pancreatic or hepatic deficiency The immediate effect of vitamin D deficiency is prevention of the calcium absorption for the deposition in bone as calcium phosphate Rickets is essentially a deficiency of vitamin D, and can be largely prevented by ingestion of fats rich in vitamins A and D Another effect of this deficiency is the inadequate ability of the ingested calcium to form soluble calcium phosphates which can diffuse from the blood through the cell membranes Subsequent precipitation of the soluble calcium from the blood into the intercellular substance of cartilage and osteoid tissue occurs under the proper alkaline pH conditions

In normal children the calcium phosphorus product in the blood is about 50 mg per cent If this product goes below 30 mg per cent, it is considered to indicate active rickets That the absorption is decreased in rickets is shown by the large quantities of calcium and phosphorus found in the feces, while only smaller quantities are excreted in the urine

1 The pathology

The outstanding feature is the disturbance of the enchondral ossification There is great irregularity and widening of the epiphyses (Fig 169) The cartilage cells accumulate but fail to differentiate The cartilage columns are not broken up, as normally, and ossification is retarded Wherever bone is formed, it is osteoid because of the absence or scarcity of available calcium Large islands of cartilage remain undifferentiated in the metaphysis, surrounded by osteoid bone lamellae (Fig 170) The microscopic picture shows irregularity of the epiphyseal plate with fibrosis of the marrow The ossification proceeds in a very irregular manner (Fig 171), and due to this irregularity the epiphyseal plate remains widened (Fig 172)

Characteristic deformations of the bone develop They consist in bowing of the long bones with such deformities as tibia vara or valga (Fig 173), and result from the irregularities in epiphyseal growth (Fig 174)

The *x ray* shows the widened epiphyseal line with irregular contours, enlarged epiphyses, and general atrophy of the bone The principal trabecular lines



Fig 170 Rickets of upper femur with coxa vara and islands of cartilage in metaphysis and neck



Fig 169 Healing rickets a) Upper humerus b) Microphotograph showing anarchic arrangement of cartilaginous cell columns irregular calcification and ossification Fibrosis of the marrow increased blood supply



Fig 172 Rickets widening of epiphyseal plate islands of cartilage in the metaphysis



Fig 173 Rachitic coxa vara

stand out prominently, and there are characteristic white, horizontal lines near the epiphysis which demonstrate seasonal fluctuation in ossification (Fig 175)

2 *The clinical pathology*

Rickets is a generalized disease, and therefore involves all systems of the body, particularly the skeletal system and the gastrointestinal tract In early rickets the head shows enlargement and incomplete ossification of the membranous preformed bone of the skull, leaving large areas of fibrous,



Fig 174 Rickets in adult irregularities of epiphyseal growth



Fig 175 X ray showing wide epiphyseal spaces with white lines in metaphysis representing seasonal fluctuations Case P S



FIG 171 Rickets of lower femur a) Microphotograph showing irregularity of the epiphyseal plate and fibrosis b) Ossification proceeding in irregular manner

(Fig 180) In these patients vitamin D is very slow in controlling the rickets, improvement is obtained by using large doses daily over an extended period of time The blood calcium is normal, but the phosphorus is diminished, blood phosphatase, blood non protein nitrogen, and the urinary output of calcium are normal

In the so called *Toni Fanconi syndrome* of rickets there is also normal blood calcium and diminished blood phosphorus, but the phosphatase is increased, and the condition is associated with polyuria, albuminuria, and glycosuria, though there is no increase in the blood sugar The process usually starts during the first two years of life, with retarded development, loss of appetite, and signs of rickets The continuous elimination of phosphorus often leads to



FIG 179 Rachitic coxa vara genu valgum and ankle valgus Case J J

hypophosphatemia and acidosis A strong alkalizing diet supported by the administration of sodium bicarbonate and lactate is being used, though the effect is doubtful Some authors claim results from vitamin D in high doses of 50,000 USP units daily On the whole, however, the prognosis is poor

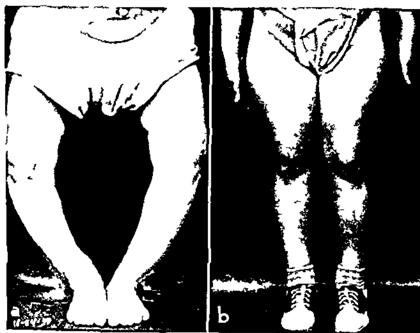


FIG 180 Resistant rickets Case R Y (female) #38 19.01 two and one half years June 1938 a) Case of resistant rickets still showing evidence of rachitic activity *after seven years of therapy b) In spite of this* operative correction of a marked bilateral genu varum (femoral and tibial) was successful and well maintained though no special precaution was taken

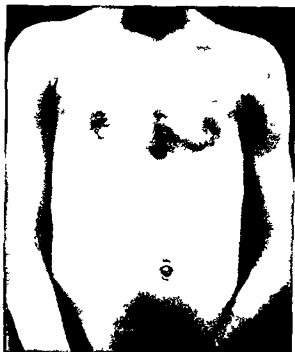


Fig 176 Rachitic pectus infundibulatum Case L E



Fig 177 Rickets protruding abdomen and depressed thorax Case C D

nonossified tissue (craniotabes) The frontal bone is protruberant The nose shows a saddle like deformity, and there is the characteristic malformation of the teeth The chest and thorax become deformed by the depression of the sternum (pectus infundibulatum, Fig 176), the ribs are slanting, and the abdomen is protruberant (Fig 177) The characteristic rachitic deformity of the spinal column is the lower dorsal or dorsolumbar scoliosis (Fig 178) which, according to Farkas, develops from a lumbar kyphosis

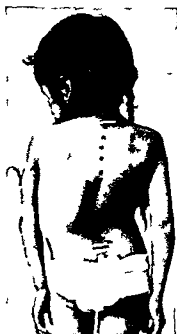


Fig 178 Rachitic scoliosis Case J A

The *upper extremities* present deformities in shape of enlarged wrist, and bowing of the long bones, especially the humerus, radius, and ulna The common *deformities of the lower extremities are* coxa vara of the upper end of the femur, bowing of femur and tibia, valgity and varity of the knee, and rachitic flatfoot and ankle valgus (Fig 179) The *pelvis* shows increased inclination, and the characteristic features of the rachitic pelvis, it is either typically flat or uniformly constricted The flatness is accentuated by the ventral projection of the sacrum, which often gives the pelvic inlet a kidney or cloverleaf shape, especially if associated with a protrusion of the acetabulum

The term *resistant rickets* is applied to cases which do not respond easily to antirachitic treatment

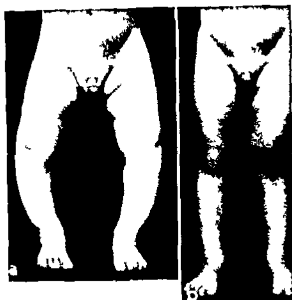


Fig 182 Rachitic bowlegs conservative treatment
a) Before b) After

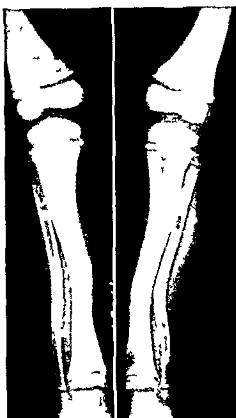


Fig 183 Rachitic bowlegs osteotomy
Case A K

these, 69 2 per cent were fully corrected Of the 21 patients treated by braces alone, 51 2 per cent were fully corrected Of the 17 cases treated by braces and shoe correction, 94 1 per cent had complete correction Thus, out of the 51 patients treated conservatively, 38 were fully corrected, six were improved, and seven failed Full correction was obtained in 74 5 per cent

2) Treatment of rachitic genu varum (G D Purvis[8]) In our clinic 179 cases were treated between 1937 and 1946 The treatment consisted in braces alone, with excellent results in 43 per cent, and good results in 48 per cent Of the cases treated by osteoclasis, 46 per cent were excellent, and 36 per cent were good results The cases treated by osteotomy showed 33 per cent excellent, and 17 per cent good results

3) Treatment of rachitic scoliosis (F J Krueger[5]) Of 20 cases treated, 16 had conservative treatment by scoliosis exercises, or exercises in combination with cast or brace, or cast only The results in these 16 cases showed four cases with fair results, seven cases with poor results, and five cases with no change observed Of the four cases which were fused, there were one good, one fair and two poor results

4) Treatment of chest deformities (F J Krueger[5]) Of 26 cases of chest deformities, 10 boys or 38 8 per cent had a pigeon chest, seven boys or 26 9 per cent, a funnel chest, five girls or 19 2 per cent had a funnel chest, and four girls or 14 per cent, a pigeon chest deformity The treatment consisted in braces or corsets and in breathing exercises While there was no remarkable change in the condition in any case, the treatment had a valuable effect on the

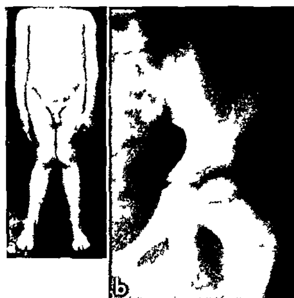


FIG 181 Renal rickets Case R N (male) #39 8085 eight years July 1939 One year ago parents noticed progressive knock knee Nine months ago child began complaining of intermittent aches in the knees and ankles Signs of rickets noted at eight months but very little response to treatment received cod liver oil and orange juice up to age of four years Patient's thorax showed Harrison's groove and rachitic rosary a) Protruberant abdomen knees in 40 degrees valgus b) X ray showed typical rachitic changes Normal calcium and phosphorus increased phosphatase and non protein nitrogen Patient died of renal insufficiency in 1945

In *renal rickets* the calcium deficiency is associated with chronic nephritis (Fig 181) The symptoms are those of rickets, with deformities of the thorax and limbs The nephrosclerosis is accompanied by decreased blood calcium, and increased phosphorus, phosphatase, and blood non protein nitrogen A similar picture is seen in cases of rickets associated with hydro ureter The kidneys show hyalinization of the glomeruli with swelling of the tubular epithelium and fibrosis, many lymphocytes and monocytes can be seen in the parenchyma of the kidney

3 The treatment of rickets

The supreme factor in the treatment of rickets is the administration of vitamin D in connection with calcium Under this treatment the blood chemistry rapidly returns to normal except in cases of resistant and renal rickets Some precaution against the toxic effect of vitamin D given in large doses is necessary As a rule, rachitic infants tolerate larger doses of vitamin D than do normal infants (Wolf[11]) However, in case of renal involvement larger doses readily result in calcium deposition Hild[4] reports a case of renal rickets in which the renal involvement was probably due to congenital malformation of the urinary tract In this case minimal amounts of vitamin D previously administered produced widespread deposition of calcium in the lungs

THE TREATMENT OF SKELETAL DEFORMITIES

The local treatment consists in braces or casts in cases up to eighteen months to two years (Fig 182) Older children two or three years of age are treated by osteoclasis, provided the bowing is in the middle of the tibia If the bowing is more in the lower portion, we prefer the osteotomy (Fig 183)

STATISTICS ON THE TREATMENT OF RACHITIC DEFORMITIES OF THE LOWER EXTREMITIES (C I NADEL[7])

1) Conservative treatment of rachitic genu valgum Of 51 cases in the series, 13 were treated for mild ankle valgus with shoe correction alone, of

osteomalacic pelvis is of the so called duck bill type, in contrast to the rachitic pelvis which is flattened in anteroposterior direction The principal feature of the osteomalacic pelvis is the deformity of the os pubis and the os ischii These bones are deflected and are bent at their weakest points around the obturator foramen In addition, the posterior portion of the os ilii is curved inward, usually behind the sacroiliac junction Due to the pushing in of the pelvis, the acetabulum points more forward, while the floor of the socket bulges out against the pelvic cavity, similar to the protrusion seen in the Otto pelvis The two horizontal rami of the os pubis are close together at the symphysis, sometimes they may touch each other, the so called symphyseal duckbill deformity



Fig 186 Osteomalacic pelvis

(W Putschar[9]) The pelvic inlet is heart shaped or clover-shaped, leaving a narrow Y shaped passage It is further narrowed by the protrusion of the fourth and fifth lumbar vertebrae (W Putschar[9]) (Fig 186)

2 Clinical signs

A *clinical* division of osteomalacia distinguishes 1 The osteomalacia due to pregnancy with an insufficient calcium intake 2 The starvation osteomalacia 3 The osteomalacia which follows idiopathic steatorrhoea

The first symptoms are pain in the back, flanks and groin, and difficulty in walking The sternum protrudes, the shoulders stoop, the trunk is bent forward, and there is the same deep transverse abdominal crease across the costal margin that one sees in rickets Spinal motion is restricted, and the anteroposterior deformity of the spine reduces the height of the patient, often by several inches Medullary compression is occasionally observed, manifested by exaggerated reflexes, clonus, and tremor The general condition of the patient

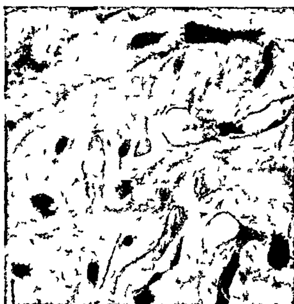


FIG 184 Osteomalacia fibrosis of marrow osteoid bone

development of the respiratory capacity and the general condition of the patient

B OSTEOMALACIA

Osteomalacia is the term given to rickets of the adult It appears at all ages in puerperal, climacteric, and senile forms, all are identical in their pathological features Osteomalacia is a disease of vitamin D deficiency, with resulting lack of calcium absorption and deposition in the bone The pathogenesis of osteomalacia is still uncertain in spite of extensive investigations, especially in the field of ovarian hormones Hypertrophy of the parathyroid was found by Erdheim and Schmorl in osteomalacic patients, but whether hyperplasia is the cause of the osteomalacia or is merely the effect of a disturbed calcium metabolism is still undecided

1 The pathology

Inasmuch as the enchondral ossification of the long bones is completed in the adult, the deficiency affects only the endosteal ossification The principal feature is that the bone absorption proceeds at the normal rate, but the apposition of new bone produces only osteoid tissue around the normal old bone trabeculae The histological picture shows fibrosis of the bone marrow, and osteoid bone is seen lining the bone trabeculae The wide meshed trabecular systems and the widening of the marrow spaces indicate an osteoporotic factor, but the main feature is the apposition of the calcium free osteoid bone, laid down by the osteoblasts of the fibrous marrow (Fig 184) The bone is not brittle as in osteoporosis, but appears extremely soft and pliable, so that it can often be cut with a knife, and therefore it undergoes the most grotesque deformations (Fig 185) The



FIG 185 Osteomalacic deformities Case J D

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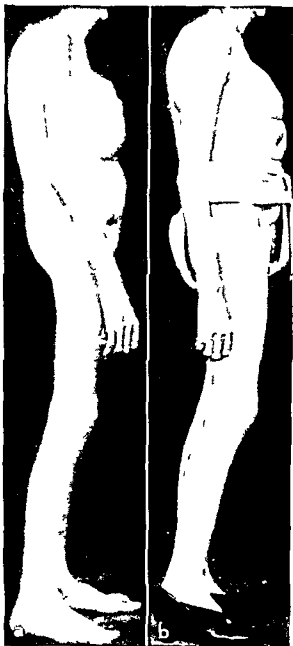


FIG 187 Osteomalacia Case M T a) Before b) After treatment

reflects the severity of the disease There is weakness, emaciation, constipation, urinary difficulty, and calculosis, besides, the patients are very susceptible to infection of the respiratory system

The *clinical, laboratory,* and *roentgenologic* findings are typical in all types There is normal calcium, diminished phosphorus, and increased phosphatase in the blood Inadequate fat absorption is manifested in some cases of osteomalacia by steatorrhoea In the case described by Miyakawa and Stearns' the patient showed a persistent elevation of the fecal fat, and the history indicated that the steatorrhoea had been present for more than ten years and may have been a major factor in the development of the osteomalacia Here the treatment required not only the administration of vitamin D in the form of cod liver oil, but also bile salts and pancreatin, and a diet rich in calcium and phosphorus (Fig 187)

Although the steatorrhoea may be symptomless and may not be associated at once with severe osteomalacia, yet a deficiency of fat absorption continued for years is very likely to lead ultimately to severe osteomalacia

3 The treatment

The treatment consists essentially in a high calcium diet and vitamin D in the form of cod liver oil or viosterol The absorption of calcium requires, besides the vitamin D, *sufficient gastric acidity and adequate hepatic and pancreatic* function to take care of the absorption of fat containing the vitamin Therefore, any deficiency in fat digestion requires the administration of bile salts and pancreatin Furthermore, in senile osteomalacia follicular hormones should be added, such as progesterone and esterone Calcium salts and parathormone are indicated in cases of osteomalacia complicated with tetany, where the calcium content of the blood is low Back pain requires immobilization in a

corset or brace Deformities of the limbs are treated according to general orthopedic principles (Fig 187)

II VITAMIN C DEFICIENCY

Cevitamic acid is essential for the integrity of the blood vessel walls Vitamin C is also of importance in the healing of wounds, and especially in bone repair It has a role in the transformation of the connective tissue into precallus in fractures Lack of vitamin C causes an arrest of the connective tissue callus and may be one reason for pseudarthrosis (Roche and Martin Poggi[10])



Fig 188 (Left) Scurvy ubperiosteal hemorrhage Case P S (female), #46676 10 months July 30 1941 Patient was well until July 1 1941 when she began crying a great deal Had a mild cold with fever of 101 degrees which persisted to the present time Developed weakness of lower extremities and tenderness on July 19 Right lower extremity showed marked diffuse swelling over lower end of right femur and knee tenderness on palpation X ray reveals subperiosteal hemorrhage lower end of femur Treatment consisted in posterior molded splint for right lower adequate diet and vitamin C End result March 1942 physical examination negative no complaints

Fig 189 (Right) Scurvy Case A G (male) #40 858? 10 months April, 1941 Full term baby twin fed on bottle and given cod liver oil since age of four months Painful swelling about the ankles for three and one half months weight loss during last one and one half months Child was malnourished and dehydrated and cried when joints were manipulated Prominent frontal bones tongue showed small hemorrhagic areas under the mucosa hemorrhage under the gums at base of teeth beading at costochondral junction Tenderness on palpation of extremities knees were swollen arm band test positive for capillary fragility Vitamin C determination 0 75 mg per cent calcium 9 4 mg per cent phosphorus 4 4 mg per cent phosphatase 5 6 Bodansky units Bleeding time 2½ minutes co coagulation time 3 minutes Wasserman negative X ray shows active scurvy Treated with vitamin C 100 mg daily two small blood transfusions End result much improved weight gain and loss of tenderness

The most outstanding example of vitamin C deficiency is *scorbutus* or *scurvy* In this disease the integrity of the blood vessels is impaired due to lesions of the endothelial cells or their cement substance Besides the circulatory system, other tissues are involved Absence of vitamin C will cause caries of the teeth, inflammation of the gums, and inclination to hemorrhage, as well as changes in the peripheral nervous system, manifested by patchy anesthesias and disturbance of reflexes

The x ray picture shows the characteristic broadening and irregularity of the epiphyseal lines The end of the metaphysis facing the plate appears drawn out and cupped Since the activity of the osteoblasts and chondroblasts is suppressed, either partly or completely, the calcified cartilage zone shows the characteristic lattice work indicating irregular calcification The subperiosteal hemorrhage casts a faint shadow starting at the lower end of the long bones but gradually extending over the entire length of the shaft (Fig 188) Sometimes oval and definite circumscribed areas of rarefaction can be seen in the epiphyseal centers of ossi



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Lecture II

ON SENILE DISEASES OF THE BONE

THERE are *only a few* pathological bone conditions peculiar to senility On the other hand, the natural somatic changes which occur in old age are of vital influence upon the course and outlook of *any disease* of the skeletal system Skeletal tuberculosis, for instance, runs a much more severe course in the aged than in the young and middle aged, and the same applies to other inflammatory conditions, to traumatic disorders, to circulatory disturbances, or the deficiency diseases which involve the skeleton The two conditions which best represent the bone pathology of advanced age are the osteoporosis and the ostitis deformans or Paget disease

I OSTEOPOROSIS

It is difficult to draw the line between senile diseases of bone and changes due to physiological involution The outstanding feature of senile bone is osteoporosis It is the result of a retardation of the natural anabolic process



FIG 190 Senile osteoporosis Case E C (female) #43 2,98 73 years March 1943 Case of senile osteoporosis following diarrhea with increased fat in stools after heavy lifting had fractures of tenth and eleventh dorsal vertebrae and sixth eighth and ninth ribs on left Treatment consisted in high calcium and vitamin diet bile salts and support of spine resulting in relief

There is a lack of bone apposition, while bone absorption goes on at a normal rate

Several factors favor senile atrophy of bone W Muller[8] observed in higher degrees of *arteriosclerosis* changes in the bone which he called fractional necrosis Although the muscles and other soft tissues are well preserved, and the bone marrow itself remains alive, areas are found in the interior of the bone which show disappearance of bone cells, empty lacunae, and necrotic bone tissue The bone structure is very sensitive to circulatory deficiency, and areas of aseptic necrosis may occur at all ages, but the precarious circulatory supply in older people with arteriosclerosis explains why those small necrotic foci occur so frequently either spontaneously or following traumatism

fication (Blitz) The earliest x ray changes are a ground glass type of demineralization of the bone structure and a signet ring appearance of the epiphysis (Evans[3]) (Fig 189) A submetaphyseal notch is seen in a large percentage of cases and, according to Evans,[3] epiphyseal displacement occurs in 19 per cent

Diagnosis Outstanding signs are general malnutrition, bleeding gums, decay of teeth, painful swelling of the long bones due to periosteal hemorrhage, swelling of the large joints, and contractures The patient seems unable to move the limbs, a so called pseudoparalysis which is due to injury of the epiphyseal plates In early or in subacute cases the method of determining scurvy is the fasting plasma determination of ascorbic acid

The *treatment* consists in giving vitamin C medicinally, and in a diet of fresh vegetables and citrus fruit containing a substantial amount of cevitamic acid Recovery is usually prompt and complete under this regime (Figs 188 and 189)

REFERENCES

1 ARAZOZA C F DE Fisiopatologia del Metabolism Oseo *Bibl del Med Practico 23* 1945 La Habana Cultural South America
2 BLITZ D *J Pediat 23* 84 1943
3 EVANS W A *Am J Roentgenol 53* 147 1945
4 HILD J R *Am J Dis Children, 63* 126 1942
5 KRUEGER F J *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *19 D 77* 1947 1948 also *19 D 75,* 1947 1948
6 MIYAKAWA G and STEARNS GENEVIEVE *J Bone & Joint Surg 24* 429 1942
7 NADEL C I Personal Communication 1947 1948
8 PURVIS G D *Research Seminar Notes,* Dept Orthop Surg State Univ of Iowa *19 D* 81 1947 1948
9 PUTSCHAR WALTER *Handb d Spez Pathol Anat u Histolog* Berlin O Lubarsch and F Henke *94* 538 1930 Fig 44
10 ROCHE J and MARTIN POGGI R *J de Chir Paris 58* 264 1941 42
11 WOLF I J *J Pediat 28* 707 1943

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REFERENCES

1 Arazoza C F de Fisiopatologia del Metabolism Oseo *Bibl del Med Practico, 23* 1945 La Habana Cultural, South America
2 Blitz D *J Pediat 23* 87, 1943
3 Evans W A *Am J Roentgenol 53* 147, 1945
4 Hild J R *Am J Dis Children 63* 126 1942
5 Krueger F J *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *19 D 77* 1947 1948, also *19 D 75* 1947 1948
6 Miyakawa G and Stearns Genevieve *J Bone & Joint Surg, 24* 429, 1942
7 Nadel C I Personal Communication 1947 1948
8 Purvis G D *Research Seminar Notes*, Dept Orthop Surg State Univ of Iowa *19 D* 81, 1947 1948
9 Putschar Walter *Handb d Spez Pathol Anat u Histolog* Berlin O Lubarsch and F Henke *94* 558 1939 Fig 44
10 Roche J and Martin Poggi R *J de Chir Paris 58* 264, 1941 42
11 Wolf I J *J Pediat, 28* 707 1943

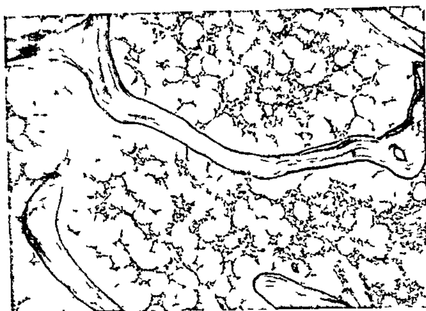


Fig 191 Osteoporosis Note atrophy of trabeculae

physiological limits the atrophy does not interfere with the normal function and reaction of bone The bone shows the same ability to form spurs and bridges in reaction to static stresses, and the same general tendency to heal when fractured

As the atrophy assumes pathological proportions under the effect of endocrine, vasomotor, or metabolic disturbances, the bone becomes more brittle and fragile, and more susceptible to fracture and subsequent deformity In the spinal column deformities occur under the influence of body weight, and are enhanced further by degenerative changes in the intervertebral discs The pressure resistance is low, and there is a great tendency to compression fractures of the vertebral bodies (Fig 192)



Fig 192 Osteoporosis of spine compression fractures Case L M

B THE CLINICAL DIAGNOSIS

The male sex prevails, the ratio being 4 1 (Black) The spine is markedly kyphotic in the dorsal section, and spinal movement is restricted The head is thrust forward, the thorax is flattened anteroposteriorly, the sternum is depressed at its base and there is a transverse abdominal crease (Fig 193) The patients complain of weakness, fatigue, and often of radiating pain The x ray shows the vertebral contours to be faint but unbroken Spurring of the edges of the deformed vertebrae, indicating reactive arthritic changes, can be noted The cortex of the long bones is greatly reduced in thickness As a result of the penetration of the disc into the vertebral bodies, the latter assume a peculiar, fishtail shape (Fig 194) Though the bones are lacking in calcium,

Defective alimentation and inanition of older people is apt to lead to loss of calcium and phosphorus and, since their excretion in the urine remains normal, the skeletal system becomes depleted Increased ingestion of calcium salts only leads to increased excretion The low gastric acidity and the hepatic and pancreatic insufficiency, frequently seen in the old, favors decalcification of the skeleton because of the resulting inability to absorb fats and, consequently, to absorb vitamin D, which is essential for the utilization of ingested calcium (Fig 190) Achlorhydria is a physiological involution, with a decline in the acidity after the age of forty to 30 35 units, the proteolytic enzymes are likewise diminished These patients suffer from lack of appetite, gastritis, and constipation and impaction

The *vitamin content* of the body is naturally low in old age The ascorbic acid falls to 0 2 0 3 mg per cent, as compared to 0 7-1 25 mg per cent normal Vitamin C is diminished in the blood serum, while lipoids and proteins are increased There is also a decrease of vitamins A and D

The *ptyalin* in the saliva is decreased, and there is incomplete digestion of carbohydrates, though to some extent the pancreatic amylase compensates for the deficiency of ptyalin amylase

Thyroid dysfunction is observed in osteoporosis, but is of more significance in senile osteomalacia Similarly, *parathyroid* dysfunction due to simple senile changes in the parathyroid gland has been reported in some cases of senile osteoporosis But it is only in true parathyroid disease, in ostitis fibrosa generalizata, where there is an excess of blood calcium and phosphatase together with a decrease of phosphorus, that the effect of hyperfunction of a parathyroid adenoma has been definitely established Some see an indication of *hypophyseal* dysfunction in the fact that osteoporosis may be accompanied by hypertension and obesity

Since *all vitamins* are necessary for the functioning of the endocrine glands, the lack of any vitamin leads to glandular dysfunction All three factors, namely, insufficiency of the glandular apparatus, lack of vitamins, and gastrointestinal dysfunction, seem so intricately linked together that it is impossible to establish the chain of cause and effect

A PATHOLOGY

Senile osteoporosis was described as a clinical entity by Charcot (1863) It is an eccentric atrophy, with coarseness of the internal architecture of the bone The haversian canals are widened The statically important trabeculae (e g, the vertical lamellae in the vertebrae) are less affected than are the secondary trabecular systems Since the porosis is due to lack of apposition only, the lacunar absorption is not increased, and the outlines of the marrow spaces remain smooth (Fig 191) Aside from the long bones and the spine, the atrophy appears in the maxillary bones and the skull In the simple senile osteoporosis there is no decalcification or production of noncalcified bone, this distinguishes it from both the rachitic and malacic changes in bone Within

Local and radiating pain is best controlled by support and physiotherapy In addition to radiant heat and gentle massage, sedatives should be given Some caution is necessary Due to the arteriosclerotic changes, the increased capillary permeability and the high blood concentration in the aged, barbiturates often show a prolonged effect

Senile Osteoporosis

Case B M (Female) #40 4280
Age 53 years Adm March 10, 1940

Patient complained of pain in the back of about one year s duration, which began when she stretched herself in bed The pain became gradually worse, and the patient has been bedfast the past several months Coughing and sneezing cause pain No abnormal dietary habits Examination showed motion of the back markedly limited The x ray revealed osteoporosis of the spine with wedging of the vertebrae Treatment consisted in bed rest, spinal support, and a high calcium, high vitamin diet

II PAGET'S DISEASE

Ostitis deformans or Paget's disease may be classed as a disease of senescence, because it occurs between the ages of forty and seventy It is about equally divided among the sexes Schmorl[12] found it in 2 75 per cent of the spines examined, and in 4 per cent of cadavers over fifty The disease derives its name from the classical treatise of Sir James Paget[10] which appeared in 1877

A THE PATHOLOGY

The principal pathological changes which Paget[10] mentions are the enormously widened haversian canals which contain a homogenous, granular substance filled with round or oval shaped cells resembling leukocytes According to

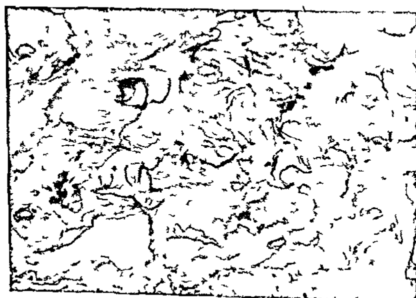


FIG 195 Paget s disease mosaics and active new bone formation





FIG 194 Senile osteoporosis



FIG 193 Senile osteoporosis vertebral collapse Case C C

there is ample evidence of calcium deposits in other structures There is sclerosis of the aorta and other vesels, calcification of costal cartilage, and calcium deposits in the digestive and urinary tracts

C THE TREATMENT

The objectives are 1) To replenish the deficiency of calcium and phos phorus in the bones 2) To prevent or correct deformities 3) To relieve inter costal pain

The *first* objective is attained by a well balanced diet, and by administration of vitamins A diet high in calcium and phosphorus should be supplemented by dicalcium phosphate and cod liver oil three times daily before meals Blood transfusions are very useful in raising the general body resistance That aged patients should be restrained in bed as little as possible to avoid hypostatic pneumonia is obvious

To *prevent deformity* of the spine use is made of corsets or, preferably, braces The spring back brace, used in our clinic, supports the spine and the anterior abdominal wall, and leaves the chest free

come more marked The bone becomes more sclerotic, hard, and brittle The fatty marrow is replaced by fibrous marrow

Pseudocysts without lining, and brown tumors are occasionally seen in the coarse trabecular pattern In the polyostotic form the skull is almost regularly involved It is hyperostotic, thickened, rough, and irregular, which gives it the so called "nigger wool" appearance in the x-ray picture (Fig 197) On the other hand, there are isolated areas of osteoporosis (Kasabach and Gutman[6]), usually starting in the region of the frontal bone, but extending into the entire calvarium Several separate foci may become confluent

The *x ray findings* of ostitis deformans in the long bones are characteristic At first, irregular areas of translucency appear, which become confluent and occupy a greater portion of the shaft The latter shows the typical bowing due to periosteal proliferation, not unlike the bowing seen in lues (Fig 198) As the bone absorption proceeds in the acute stage, the whole shaft takes on a coarse, masked texture while the bowing progresses When the periosteal reaction lags behind while the endosteal absorption proceeds, the whole shaft assumes a bubble like, blown up appearance (Fig 199) However, there is al



Fig 198 (Left) a b Paget s diseas bowing of the shaft
Fig 199 (Right) Paget s disease of the tibia bubble like appearance of shaft Case A F



FIG 196 Paget s disease a) Active periosteal new bone formation b) Active osteoclasis

newer concepts (Schmorl[1]), the disease does not really begin at the bone lamellae themselves, but in the bone marrow, with a loosening of the endosteum followed by catabolism of the lamellae It is believed (Koops[7]) to be a creeping serous inflammation which in the marrow leads to fibrosis, and in the haversian system to the formation of new bone laid down in a peculiar mosaic like pattern characteristic of the condition The first changes then consist in this loosening of the endosteum, then the absorption of bone, the fibrosis of the marrow and, finally, the new bone formation and apposition in "mosaics" (Fig 195) These changes represent three different stages

1 The acute stage is marked by pronounced hyperemia and rapid bone absorption The marrow is replaced by young fibrous tissue New bone trabeculae are formed from the periosteum, as well as from the endosteum Mosaic structures begin to appear The bone becomes thicker, and the periosteal fibrous bone becomes mature The endosteal bone apposition produces a narrowing of the medullary spaces (Fig 196)

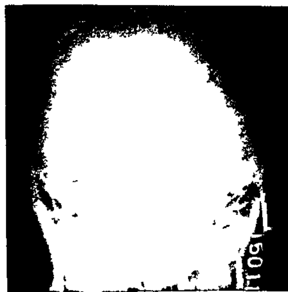


FIG 197 Paget s disease involvement of skull

2 Later the osteoblastic activity holds the balance to the earlier osteoclastic activity More and more of the fibrous bone is replaced by mature Paget bone

3 In the third or healing stage the osteoblastic activity preponderates The mosaic structures be

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Fig 198 (Left) a b Paget s disease bowing of the shaft
Fig 199 (Right) Paget s disease of the tibia bubble like appearance of haft Case A F



Fig 200 Paget s disease stages of absorption and healing Case V C

most always a sharp demarcation from sound bone, usually in the form of a V-shaped line which, when present, is almost pathognomonic In the later stage, when bone apposition prevails, the shadows become more dense and homogenous (Fig 200) Involvement of the patella is occasionally observed (Fig 201)

In 90 per cent of the cases of Paget's disease the spinal column is involved Most commonly affected are the middorsal and the lumbar spine, the cervical section remains free In the spine a malacic or osteoporotic phase also precedes that of condensation In the early stages there is rarefaction and coarse trabeculation Then follows a stage of progressive condensation and opacity and sharpness of contours These phases may be seen concur



Fig 201 (Left) Paget s disease of the patella. Case R W (female) 2 years Pain and swelling of left knee for one year following a minor trauma A large patella (2½" x 2") was removed at operation the joint cartilage was slightly degenerated Slide showed fibrous marrow and abundant new bone formation and many osteoclasts A mosaic structure was present in many areas of the bone

Fig 0? (Right) Paget s disease of the spine Note flattening Case D N L



Fig 203 Paget s disease of the pelvis Case G M

rently in different portions of bone, and the x ray may show rarefaction as well as condensation Under weight bearing the Paget vertebra flattens, and this is followed often by reactive changes resembling those of osteoarthritis (Fig 202)

Breus and Kolisko[3] first described the so called *Paget pelvis* It shows the characteristic structural changes already mentioned in the long bones, and also certain deformations reminiscent of those seen in rickets and osteomalacia The lateral walls of the pelvis are flattened, and the acetabulum protrudes slightly into the pelvic cavity, causing a narrowing of the pelvis from side to side, and a duck bill deformation of the symphysis (Fig 203)

A more unusual location of Paget's disease is in the long bones of the upper extremity Characteristic bowing occurs, similar to that in the weight bearing bones (Fig 204)



Fig 204 Paget s disease of the radius progressive deformity Phosphatase 24 8 Bodansky units X ray showed typical Paget s and was confirmed by biopsy Case N C

B THE CLINICAL COURSE

Paget's disease occurs in monostotic and polyostotic forms The monostotic type is the more common, it is confined to a circumscribed area in one bone, most often in the tibiae, the vertebrae, and the sacrum The polyostotic in

volves a number of bones including both tibiae, the vertebrae, and almost always the skull

The principal clinical signs are 1) Pain, which is dull, boring, and osteocopic, and 2) deformation of the long bones of the lower extremity, of the tibia and femur, with secondary static disability of the knees If the *lower spine* is involved, the normal lumbar lordosis is effaced, and mobility is much restricted Due to hypertrophic cranial changes, the size of the skull increases

The *blood analyses* show normal calcium, low phosphorus, and increased alkaline phosphatase This increase often reaches 100 to 160 Bodansky units, and is very suggestive of Paget's disease It is by no means constant, however, because it depends on the bone activity, and hence may disappear in stages of quiescence Besides, there are other bone conditions, such as osteoporosis, renal rickets osteomalacia, multiple myeloma, and metastatic carcinoma, which may show increased phosphatase in their active stages

C COMPLICATIONS

1 Spontaneous fractures in Paget's disease

Pathological fractures are rare in Paget's disease They seem to occur both during the active stage and during the healing stage Fissure fractures of the cortex (Allen and John[1]) may be produced by slight traumatism, and may go on to complete transverse fractures There is usually absence of pain The callus appears early (Roger and Ulin[11]) (Fig 205), and it assumes the histological characteristics of Paget's disease

2 Spinal compression in Paget's disease

Compression myelitis is occasionally observed in Paget's disease of the spine The cause may be a direct pressure from a collapsed vertebra or some hypertrophic bone formation, or it may be a concomitant vascular lesion which produces sclerosis or malacia of the cord A transitory quadriplegia and a persistent Brown Sequard syndrome following sudden collapse of a Paget vertebra is reported by Garcin in a patient with involvement of the cervical spine Simple compression paraplegia with spastic paralysis, sphincter and sensory disturbances, signs of spinal blockage, increased globulin, progressive weakness and numbness in both legs, atrophy, and absence of reflexes has been observed Cord lesions occur most frequently in the dorsal spine

3 Sarcoma in Paget's disease

In about 10 per cent of the cases of polyostotic Paget's disease sarcomatous degeneration occurs It is claimed that mechanical stress (Davie and Cooke[4]) and preceding trauma predisposes to malignant degeneration (Fig 206)

D THE TREATMENT

There is no specific treatment for Paget s disease The symptomatic treatment is directed mainly toward the control of pain



FIG 205 (Left) Paget s disease of the femur pathologic fracture Case E A

FIG 206 (Right) Paget s disease of the right femur with sarcomatous degeneration Case C M (male) #43 5534 47 years November 1943 Patient entered complaining of pain in right thigh for last 10 years associated with bowing of the right femur three to four months ago he suddenly began having severe boring constant pain in the right thigh X ray showed fairly far advanced Paget s disease of the right femur Biopsy showed Paget s disease with sarcomatous degeneration (giant cell or osteolytic type) Treatment consisted in stilbestrol with relief for three weeks and then return of pain Patient died 11 months later

1 Medicinal

Iodides, arsenic, and endocrine therapy are of doubtful value Lately, aluminum acetate (aluminum acetate ¼ fluid ounce, syrup 300 minims, essence of cherry 4 minims, mel depuratum ad 4 ounces, minimum dose 1 fluid ounce four times daily after meals, together with 1 pint of milk daily) has been advocated on the supposition that it reduces absorption of phosphorus According to a report of Helfet,[5] eight cases so treated responded with loss of pain and fatigue, and gain in weight

2 Physiotherapy

X ray treatment gives temporary relief in some cases It is worth trying before surgical intervention is resorted to *Ultraviolet rays* have occasionally

given encouraging results (Nicory), and *heat* in one form or another may afford relief

3 Surgical

Robert Jones recommended periosteal stripping for the relief of pain Drilling or windowing of the bone alleviates pain by releasing the intra osseous pressure, as we have observed in some cases, the relief, though temporary, may last for a number of months to a year This procedure requires a great deal of caution, because the bleeding from the bone may be profuse and difficult to control

In spinal compression, with paraplegia or spasticity, decompression (laminectomy) has been carried out with success Schwartz and Robock[13] reported three cases, of these, one died and two recovered from the paraplegia Of five other cases quoted in the literature, one died and four recovered

REFERENCES

1 ALLEN N L and JOHN R L *Am J Roentgenol* July 1937
2 BLACK J R *Proc Staff Meet Mayo Clin* *15* 39 619 Sept 15 1940
3 BREUS C and KOLISKO A Die Pathologischen Beckenformen Wien und Leipzig F Deuticke 1900-1911
4 DAVIE T B and COOKE W E *Brit J Surg* *25* 299 Oct 1937
5 HELFET A J *Brit J Surg* *27* 651 April 1940
6 KASABACH H H and GUTMAN A B *Am J Roentgenol* March 1937
7 KOOPS H *Virchows Arch f path Anat* *304* 297 1939
8 MULLER W *Normale und Pathologische Physiologie des Knochens* Leipzig 1924
9 NICORY C *Brit M J* *1* 479 March 15 1930
10 PAGET SIR JAMES *Med & Chir Trans* *60* 37 1877
11 ROGER M H and ULIN R *J Bone & Joint Surg* Oct 1930
12 SCHMORL G *Virchows Arch f path Anat*, *285* 694 1932
13 SCHWARTZ G A and ROBOCK S *Am J Roentgenol* *42* 545 Sept, 1939

Lecture III

ON DEGENERATIVE BONE DISEASES

I. OSTEITIS FIBROSIS CYSTICA (LOCALISATA)

A. ORIENTATION

THE localized fibrocystic disease of bone, though not a disease of advanced age, may be classified among the degenerative bone diseases. As the name implies, its constant feature is the fibrous transformation of bone marrow. This in itself is not characteristic for the disease, but is merely a nonspecific reaction of the bone marrow to some hitherto unknown agent. We observe the same phenomenon in the generalized osteitis fibrosa, or Recklinghausen's disease, in which we know that it is an endocrine disturbance due to dysfunction of the parathyroid gland. We see it in Paget's disease, a condition of entirely unknown etiology. In the osteitis fibrosa of Recklinghausen bone absorption is rapid and progressive. In Paget's disease the osteolytic phase is followed by a period of bone production, however irregular and erratic. We further note fibrosis of the marrow in simple osteoporosis and in the malacic diseases of rickets and osteomalacia. Albright's disease, a condition associated with pathological pigmentation and endocrine dysfunction, is likewise characterized by fibrosis of the marrow, not to speak of the infectious diseases in which medullary fibrosis is a common feature of inflammatory reaction.

It seems strange, therefore, to set up such a common occurrence as marrow fibrosis as the criterion for a distinct pathological entity. Still, in osteitis fibrosa cystica localisata, or the fibrous dysplasia as it is called, the fibrosis of bone marrow is such a dominant feature, and other pathological phenomena seem so secondary to it, that for lack of more precise knowledge of its pathogenesis this condition must remain classified under the general term of degenerative bone disease.

The term polyostotic fibrous dysplasia was first suggested by Lichtenstein[15] for a disease that affects several bones with a tendency to unilaterality.

B. THE PATHOLOGY

The lesion is usually multiple, although there are many observations of solitary lesions. Albright's disease seems to be a variant, characterized by pigmentary skin changes and precocious puberty. The lesion presents a fusiform, symmetrical enlargement of the bone with thinning of the cortex. The cancellous bone is replaced by a firm, yellowish white tissue which contains small cysts filled with fluid. The principal component of the lesion is connective tissue, arranged in bundles within which trabeculae of newly formed bone are found (Schlumberger[3]) (Fig. 207). The bone is metaplastic and formed from

given encouraging results (Nicory[9]), and *heat* in one form or another may afford relief

3 Surgical

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5 Helfet A J *Brit J Surg, 27* 651 April 1940
6 Kasabach H H and Gutman, A B *Am J Roentgenol* March 1937
7 Koops H *Virchows Arch f path Anat 304* 297 1939
8 Muller W *Normale und Pathologische Physiologie des Knochens* Leipzig 1924
9 Nicory C *Brit M J 1* 479 March 15 1930
10 Paget Sir James *Med & Chir Trans 60* 37 1877
11 Roger M H and Ulin R *J Bone & Joint Surg* Oct, 1939
12 Schmorl G *Virchows Arch f path Anat 283* 694 1932
13 Schwartz G A and Robock S *Am J Roentgenol 42* 345 Sept 1939

specific, and it is difficult to differentiate from cysts, giant cell tumors, chondromas, and fibromas

D THE CLINICAL SIGNS

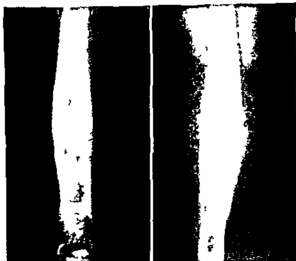


FIG 210 Osteitis fibrosa cystica expansion of shaft Case L F

The initial sign is local swelling and tenderness In the monostotic form the swelling is usually around the shaft of the tibia, while in bone cysts the preferred site is the upper end of the tibia and femur Pathological fractures are not uncommon (Fig 211) They heal promptly on immobilization In many cases the disease is confined to one bone alone, osteitis fibrosa monostotica (Fig 212) Unilateral cases were reported from our clinic by Freund[11] Involvement of the vertebral column is rare One case involving the cervical region is described by Schlumberger[3] There were signs of local tenderness and radiation down the arm The ribs are involved in over 40 per cent of the cases



FIG 211 Osteitis fibrosa localisata (monomelica) pathological fracture



FIG 212 Osteitis fibrosa cystica monostotica




FIG 207 (Left) Osteitis fibrosa cystica localisata fibrosed marrow with new formed bone trabeculae
FIG 208 (Right) Osteitis fibrosa cystica localisata large expanding cyst in the metaphysis Case E L

connective tissue The connective tissue cells become more vesicular and, being surrounded by increasing amounts of calcifying intercellular substance and attached to metaplastic bone, they become indistinguishable from osteoblasts In close proximity to these osteoblasts one finds large multinucleated cells which act as osteoclasts, breaking down the newly formed bone (Schlumberger [3]) A circle is thus formed between osteoblastic and osteoclastic activity in which, however, the latter prevails There are other large areas of fibrous dysplasia in which no osteoblastic activity nor any metaplastic new bone formation can be seen Here the changes remain prevailingly osteolytic



C THE X RAY FINDINGS

It seems that the first changes take place in the metaphyses of the long bones, areas of translucency appear which soon become confluent The process then extends into the diaphysis and often involves the entire shaft (Figs 208 and 209), delicate trabeculae traverse the areas of translucency The cortex is very thin and is expanded by periosteal bone apposition (Fig 210) The ribs show a blown up appearance with wide meshed, coarse trabeculation On the whole, the x ray appearance has nothing



FIG 209 Osteitis fibrosa cystica involving entire shaft Case J B

generalisata, or Recklinghausen's disease Some confusion arose fom the fact that he considered the fibrosis a specific feature, whereas we know today that fibrosis of the marrow occurs also in rickets, osteomalacia, and Paget's disease He also believed that the cystic expansion and the formation of giant cells were signs of malignancy (multiple osteosarcomas), while we now recognize that infiltrating growth does not prove the malign character, and that the giant cell tumors occurring in Recklinghausen's disease are biologically benign

The first light on the pathogenesis of the disease came in a rather indirect way from the observations of Erdheim[8] He found that in osteomalacia a

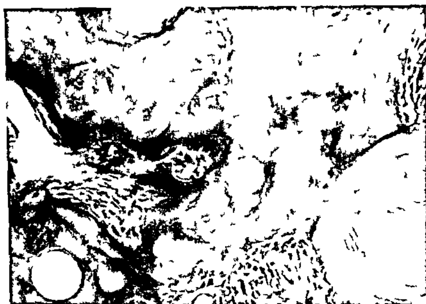


Fig 214 Recklinghausen's disease osteoclastic activity of fibrosed marrow

hyperplasia of the parathyroid may be present, which he thought to be secondary to the abnormally large amounts of calcium and phosphorus liberated from the decalcified skeleton His finding of decalcification in connection with parathyroid hypertrophy only partially cleared the situation, because he considered the parathyroid changes as secondary, and the bone absorption as primary Even before him, Askenazy[2] (1903) had published autopsy findings of generalized decalcification of the skeleton in connection with a parathyroid tumor Since then the number of reports increased steadily, reaching 75 cases by 1930 (Barr, Bulger and Dixon[3])

The credit for establishing clinical proof of the connection between the parathyroid and osteitis fibrosa belongs to F Mandl[18] He removed a parathyroid tumor, reasoning that the hypertrophy of the gland is the cause, not the effect (Erdheim[2]), of the bone disease Removal of the gland resulted in a striking improvement which lasted five years

B THE PATHOLOGY

Recklinghausen[1] in his description of the fibrous transformation of the

E THE TREATMENT

It depends much on the site and extent of the lesion Simple excision suffices for the involved ribs Curettement of the bone and filling with bone chips is the standard treatment for the long bones (Fig 213) The prognosis is good in localized cases where a thorough curettement can be done, and where there is ample bone available to fill the cavities It is more doubtful in later stages when a large portion of the shaft has become affected Recurrences are frequent In



Fig 213 Osteitis fibrosa cystica treated with curettement and bone chips Case F L a) Before b) After treatment

many cases the original appearance is one of a simple bone cyst, but following operation an extension of the condition toward the shaft is noted It seems that the degenerative process exhausts itself, but not before the greater portion of the bone has become involved Following operation protection by braces is required for a long time, especially in the polyostotic or unilateral type[5]

II OSTEITIS FIBROSA GENERALISATA (PRIMARY HYPERPARATHYROIDISM)

A ORIENTATION

Many observers have described general decalcification of the skeleton with formation of cystlike cavities, resorption of bony tissue, and replacement by fibrous tissue It fell to Recklinghausen (1891) to give the condition its precise pathological distinction as a fibrous dysplasia involving all the bones of the skeleton The disease is since known under the name osteitis fibrosa

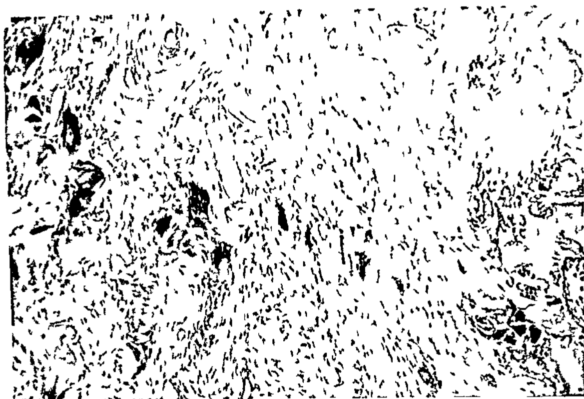


Fig 216 Recklinghausen's disease decalcification and osteoclasis

C THE BIOCHEMISTRY

There is an increase of the *blood calcium* in all cases, even though it may not be found consistently during the entire course In some of our cases it reached as high as 19 mg per cent, and even higher readings are reported The *urinary calcium* often but not always exceeds the normal values by 400 500 mg per cent The excretion of abnormal amounts of calcium in the urine causes its supersaturation with calcium salts, especially calcium phosphates Renal stones are frequent, and produce severe kidney damage and uremia Besides, toxic necrosis of the kidney may result from excess parathyroid hormone in acute exacerbations of hyperparathyroidism Fibrosis and vascular changes are found in the kidneys

The *inorganic phosphorus* in the blood is decreased *Phosphatase,* a ferment liberating inorganic phosphorus from organic compounds, is increased as in all bone diseases in which there is active new formation of bone or cartilage (Paget's, rickets, and osteomalacia)

The *parathyroid hormone* has a specific stimulating action on osteoclasts, causing widespread absorption Fibrosis of the marrow is secondary The hormone also affects phosphate metabolism by lowering the renal threshold for phosphorus excretion, thus increasing the phosphorus output through the urine Fibrous osteitis can be produced experimentally in guinea pigs by excessive amounts of parathyroid hormone However, the fibrosis of the marrow is not specific for the action of this hormone The same effect of osteoporosis can be

marrow called attention to the softness of the skeleton, the spontaneous fractures, and the friable, deformed bone which could be cut with a knife He distinguished it from osteomalacia on the basis of the fibrosis, the bone cysts, and the brown tumors, and he emphasized the presence of giant cells The gross pathology shows multilocular cysts lined by a thick wall of fibrous tissue (Fig 214) The bone is extremely soft, and shows the most grotesque deformities

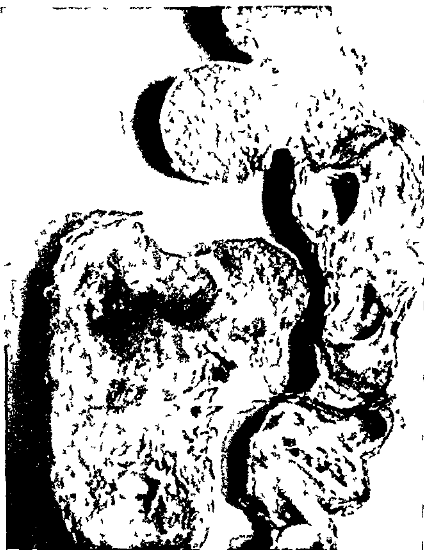


Fig 215 Recklinghausen's disease extreme deformities in cystic degenerated femur Museum specimen

(Fig 215) Microscopically one sees typical fibrous marrow, especially around the trabeculae (Fig 216) There are numerous active osteoblasts and osteoclasts Large areas may be seen entirely devoid of bone trabeculae, but containing mixed fatty and hematogenous marrow and diffuse hemorrhage These are the so called brown tumors The characteristics of Recklinghausen's disease are 1) Osteoclastic destruction of trabeculae 2) Decalcification of remaining trabeculae 3) Proliferation of fibrous tissue in bone marrow 4) Thinning and perforation of the cortex

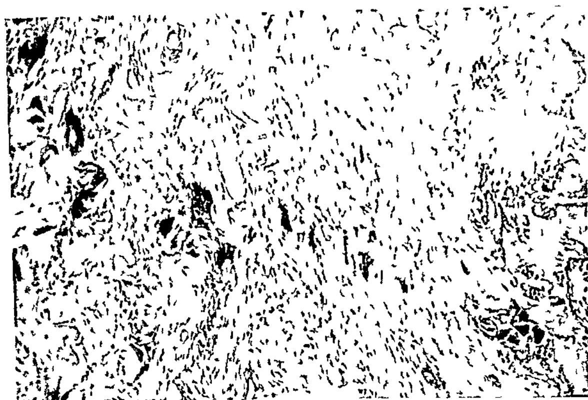


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obtained from lead acetate, chloroform, ammonium chloride, and glucose, in fact, any experimentally produced *acidosis* results in bone lesions which closely resemble fibrous osteitis It can also be produced in chickens by a vitamin D deficient diet

D THE X RAY FINDINGS IN RECKLINGHAUSEN'S DISEASE

The marrow spaces are wide, the cortex is thinned, and the haversian canals are enlarged (Figs 217 and 218) The skull has a stippled appearance (Fig 219) Cystlike areas of decalcification are seen in the wrist and fingers



Fig 217 (Left) Recklinghausen's disease wide marrow spaces and thin cortex Case J W

Fig 218 (Right) Recklinghausen's disease wide marrow spaces and thin cortex Case J W

E THE PRINCIPAL CLINICAL SIGNS

The principal *clinical signs* are as follows

1 *Muscular hypotonia*

The calcium affects the tonicity of the muscles and decreases their excitability In contrast, the excitability of the muscles is increased in hypocalcemia, and tetany results The hypotonia of the muscle leads to early fatigue

2 *Pain and tenderness*

Spontaneous pain is a frequent complaint The entire skeleton is very sensitive to touch and pressure

3 *Gastrointestinal disturbances*

Not uncommon is uncontrollable vomiting It appears early, sometimes even in the initial stage before the skeletal changes become marked

4 *Oliguria*

A temporary *oliguria* must be expected in cases of urinary calcinosis, especially if there is already some kidney damage

5 *Cachexia*

Cachexia is observed in advanced cases In this stage there is usually considerable impairment of function of the kidney and the heart, and a general edema appears

F THE TREATMENT

1 Conservative

So far all attempts to control the disease by conservative means have been of doubtful value In some instances it has been reported that calcium given in large doses, and especially a high phosphorus intake, had a favorable influence, but high calcium ingestion favors urinary calcinosis X ray treatment has been tried, occasionally with good results (Weil [8]), although the normal parathyroid gland is not radiosensitive

2 Operative

Since Mandl discovered the relationship between osteitis fibrosa generalisata and parathyroid hypertrophy, the removal of the gland has become the standard treatment Sometimes, but not often, the enlarged gland can be felt



FIG 219 (Left) Hyperparathyroidism characteristic stippled appearance of skull Case D L
FIG 220 (Right) Osteitis fibrosa cystica generalisata Death after hyperparathyroidectomy due to cachexia and kidney damage Case A W

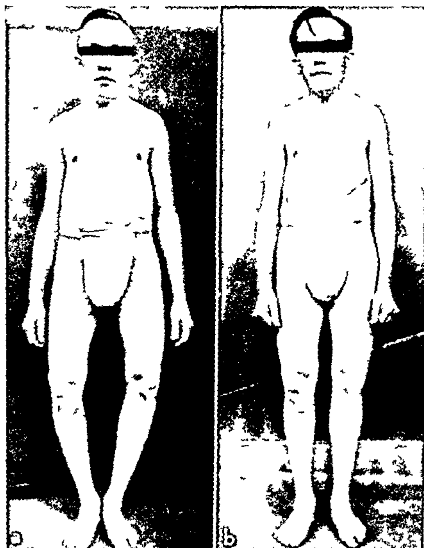


Fig 221 Recklinghausen's disease parathyroidectomy and osteotomy of tibiae Case M F a) 1929 before operation b) 1931 after operation

(Snapper) If the parathyroid tumor is calcified, as occasionally happens, it may be visualized in the x ray (Bergstrand[4]) Snapper[26] advises to look for the superior parathyroid gland at the level of the cricoid cartilage, and for the inferior at the inferior pole of the thyroid If no parathyroid tumor is found there, it may be ectopic even behind the sternum, or, worse still it may be incorporated in the thyroid gland and lie within a duplication of the thyroid capsule

The *results of parathyroidectomy* are very favorable, provided there is no great kidney damage, and the patient at the time of operation is still in satisfactory general condition One case, a 54 year old woman (A W) succumbed after operation from kidney complications and cachexia (Fig 220) Another patient (M F Fig 221) 19 years of age with a blood calcium of 19 mg per cent and a large parathyroid adenoma maintained his improvement during 11 years of observation, although parathyroid hormone therapy became necessary from time to time when the blood calcium fell too low

THE POSTOPERATIVE HYPOCALCEMIA AND TETANUS

The decalcified skeleton absorbs calcium from plasma so greedily that the calcium current which before operation was from bone to blood is now reversed, and even the remaining parathyroid gland cannot stop it Intravenous injections of calcium chloride, together with daily injections of 100 units of parathormone may become necessary In one of our cases (M F) tetanus developed almost immediately after operation, and calcium therapy with parathormone had to be instituted

III HYPERPLASIA OF PARATHYROIDS SECONDARY TO OTHER BONE DISEASES

In 1907 Erdheim[9] observed hyperplasia of the parathyroids in osteomalacia Later he[10] found it in rickets and experimental rickets It was also found in cases of multiple myeloma (Barr, Bulgar, and Dixon[3]) In chickens avitaminosis D leads to hyperplasia of the parathyroid A compensatory hyperplasia of the parathyroids associated with decalcification of bone and osteitis fibrosa is seen also in patients with steatorrhoea or biliary fistula (Parlan, 1905)

In renal rickets (congenital anomalies of the kidneys or ducts), renal osteodystrophy, or renal osteofibrosis, several authors have emphasized the occurrence of parathyroid hyperplasia (renal hyperparathyroidism)

In adults with renal failure of long duration we see calcium deposits near the joints, extreme calcification of the media of the arteries, generalized osteitis fibrosa, and enlargement of the parathyroids It is probable that the hyperphosphatemia or the hypocalcemia acts as a stimulus for the parathyroids According to Albright, *et al*,[1] a *single* parathyroid adenoma is in favor of primary hyperparathyroidism, while *multiple* parathyroid adenomas consisting of water clear cells are in favor of secondary parathyroidism Marked hypercalcemia indicates primary, and congenital renal anomalies secondary hyperparathyroidism

Why does renal failure sometimes result in rickets, sometimes osteitis fibrosa, and sometimes osteoporosis? According to Pappenheimer,[19] it depends on the calcium content of the blood, a low calcium content of the blood results in infantile rickets, a slightly higher calcium content in the presence of renal insufficiency leads to a combination of rickets and osteitis fibrosa

IV SUMMARY

1 Parathyroid adenoma is the cause of Recklinghausen disease (*primary* adenoma)

2 *Secondary* hyperplasia develops as a result of other diseases with extensive decalcification (Erdheim)

3 *Experimental* hyperplasia has been obtained in chickens by avitaminosis D, in dogs by biliary fistulas, and in rats by chronic renal insufficiency

4 *Secondary* hyperplasia is found clinically in osteomalacia, multiple mye-

lomas and malignant metastases, and in chronic renal failure with acidosis in the form of osteoporosis, renal rickets, and osteitis fibrosa

V LIPOID DYSTROPHIES [7]

The lipoid dystrophies are disturbances of the general fat and lipoid metabolism The fat resorption through the intestines is mediated by the action of bile salts, under the physical laws of diffusion, and is controlled by hormones The sustaining hormone is that of the adrenals This group of dystrophies includes such conditions as Hand Schuller Christian disease, Letterer-Siwe disease, Gaucher disease, and the eosinophilic granulomas In all of these the real cause remains obscure, but more recent pathological studies seem to indicate that they all represent variants of the same underlying disturbance of lipoid metabolism



FIG 222 Eosinophilic granuloma showing eosinophiles giant cells, mononuclears lymphocytes and fibroblasts Case M M

A EOSINOPHILIC GRANULOMA

This is a rare, benign condition of the bone of unknown etiology The term was proposed by Jaffe and Lichtenstein (1940) for what they considered a new destructive disease entity in bone At present about 30 cases of this tumor have been reported in the literature, and the vast majority were solitary lesions The most frequently involved bones are the skull and ribs, though any bone may be the seat of the disease The sequence according to frequency is skull, ribs, long bones and scapula

It is a soft, pliable tumor of purplish color, which thins out the bony cortex by absorption The soft tissue can be readily separated from the bone, leaving a smooth surface Microscopically, this granuloma presents a varied appear-

ance The essential structure is a cellular, poorly vasculnrized mnss of large mononucleir cells with i greit mny eosinophils, lirge giant cells, fibroblists, and snill mononucleir cells and lymphocyes There is evidence of hemorrhage, old or recent, nnd of lirge mononucleir phigocytes laden with hemosiderin The most striking fentures are the lirge gint cells which vary in size from 30 to 60 microns ind hnve mostly single lirge, round, ovil, or cleft nuclei The eosinophils are very numerous Mitotic figures are fairly frequent (Fig 222) Occasionally osteoblistic new bone formation ind extensive old and recent hemorrhages are observed



FIG 2?3 Eosinophilic granuloma Case A H (male) #47 517 46 years January 8 194? Patient was kicked in the left tibia one year ago three weeks later the knee became swollen Bloody fluid was obtained by aspiration Pain in the knee increased slowly and the patient imped two weeks ago pain became excruciat ing and forced him to use crutches There was infiltration of the soft tissues on the lateral a pect of the thigh knee motion was normal Biopsy showed a grape size defect in the femu filled with a dark hemorrhagic and semigelati nous material hi tological examination re vealed eosinophilic granuloma Patient us ta ned a pathological fracture while turning in bed (Fig 2?5) Treatment consisted in curet tage of tumor and insertion of bone chips but tumor recurred X ray treatment was followed by *amputation* Patient was observed for six years and had no recurrence

1 Diagnosis

Most patients reported were young male adults or children Only three were over 20 years of age The usual clinical signs are pain and swelling Sometimes a preceding trauma is reported (Case A H) The swelling increases steadily, and the neighboring joint (knee or elbow) is held rigid, or its motion is at least restricted With increasing tumor growth pain may become excruciating Weight lost is often observed Aspiration sometimes yields a turbid, blood tinged fluid Cultures are negative The patients usually show no signs of general illness, even those with multiple foci One such case with foci in both femora, both tibiae, both iliac bones, several ribs, clavicle, ulna and radius was observed by us for a number of years

X Ray Findings

The x ray shows irregular, multilocular, osteolytic areas in the shaft with little or no periosteal reaction (Figs 223, 224 and 225)



Fig 224 Eosinophilic granuloma of humerus Case M M (male) #39 387 16 years August 1938 Cyst in lower end of left humerus with pain and inability to fully extend the elbow for past three months Curettement of cyst and insertion of bone chips Pathological specimen shows eosinophilic granuloma No recurrence Four years later a cystic defect appeared in the left femur and skull and was treated by x ray Lesions in the skull humerus and femur all healed Ob ervation four years



Fig 225 Eosinophilic granu loma pathological fracture Case A H

2 Treatment

It is generally advised to treat the condition by excising the cystic areas and filling the cavity with bone chips, and then using radiation therapy The x ray dosage recommended by Green and Farber is a total of 1500 R Amputation is seldom indicated It became necessary in one of our cases (A H) because of local recurrence

B HAND SCHÜLLER CHRISTIAN DISEASE

The separation of the solitary eosinophilic granuloma from other lipoid con ditions is made on clinical grounds only Pathologically they all belong to the same group of lipoid granulomatoses In *Hand Schuller Christian* disease we find multiple, round defects in the bones of the skull, exophthalmus, diabetes insipidus, and infantilism Hand,[13] who described the first case (1893), thought the condition to be tuberculosis Schuller [4] recognized it as a pituitary dysfunc tion, and Christian's[6] monograph on the subject brought the condition to general attention

The disease is essentially a lipoid granuloma located preferentially in the skull, where it produces areas of absorption (Fig 226) The irregular, map like, serpentine absorption areas are known as "geographical" skull The disease also attacks the orbit, and the retro orbital granuloma produces the exophthalmus It also involves other bones, such as the mandible, where it causes looseness of the teeth, or of the temporal bone, resulting in deafness



Fig 226 Hand Schuller Christian disease geographical skull Case B R

Outside the skeletal system it produces eczema, and xanthelasma-like skin eruptions It involves the liver, spleen, and kidneys with xanthomatous granulations It is not a xanthoma, but a granulomatous formation in which large areas of xanthoma cells are found The principal histological feature is the large number of cholesterol containing foam cells Rowland believed the condition to be a primary disturbance of the lipoid metabolism, which links it with Gaucher and Niemann Pick diseases However, the nature of the lipoid deposited in these cells is different It is kerasin in Gaucher, phosphatides in Niemann Pick, and cholesterol in Hand Schuller Christian disease

C NIEMANN-PICK DISEASE'

Niemann Pick disease is a disturbance of the lipoid metabolism by which the cells of the central nervous system are flooded with phosphatides Occasionally familial, it affects chiefly the Jewish race (50 per cent), it is confined to children The female sex prevails

The principal signs are enlargement of the liver and spleen, brown pigmentation of the skin, enlargement of the lymph glands, febrile attacks, and amaurotic idiocy

The pathological basis is the deposition of a lipoid substance (phosphatides) into the cell protoplasm, leading to swelling of the cells The changes in the liver, spleen, adrenals, kidneys, lymph glands, thymus, and lungs consist in the formation of round or polygonal cells, in which the cytoplasm is vascuolized, owing to deposition of lipoids, so called foam cells These may replace the normal cells entirely

In the eye ground there is a greenish gray ring around the fovea, due to edema Masses of foam cells are found in the bone marrow The lymph glands are enlarged The blood shows leukocytosis Hypercholesterinemia is common

Niemann Pick disease is primarily mesodermal, and attacks the endo- and ectodermal tissues of the brain secondarily

D LETTERER SIWE DISEASE"

This condition is a reticulo endotheliosis The lesions are widely distributed through the body, including the skeleton (Fig 227) This disease appears in

infants and children below the age of two years, and is usually fatal within a few months Here the principal findings are fever, enlargement of the spleen, liver, and superficial lymph glands, and a cutaneous eruption such as purpura, together with destructive lesions of the bones, particularly of the skull (Fig 226) When the disease has run for more than one year, the histiocytes become lipophagic foam cells, similarly as in Hand Schuller Christian disease

E GAUCHER DISEASE

Gaucher[12] (1882) described a case of idiopathic enlargement of the spleen in which the pulp was completely replaced by large pale cells, at first thought to be malignant Not until 1916 was it suggested by Mandelbaum and Downey[17] that the disorder was one of lipoid metabolism, later Epstein[7] showed that the lipoid is a complex cerebroside, called kerasin



FIG 227 Letterer Siwe dis ea e combination irregular bone destruction and regional atrophy localized Ca_e J M

The spleen is enlarged, firm, and tough, its cells are replaced by large foam cells containing kerasin The bone marrow is also invaded with these Gaucher cells, causing atrophy and destruction The lymph nodes are enlarged, and are filled with Gaucher cells

The x ray shows moth eaten areas, especially in the femur and ribs (Fig 227) The site of pre dilection is the lower end of the femur, which often appears blown up and bell shaped (Erlenmeyer flask shape) Otherwise the bone shows swelling, decalcifi cation, and blurring of its structure

The disease is familial, and involves several mem bers of the generation It begins in infancy, and progresses slowly With the gradual enlargement of the spleen cutaneous hemorrhages appear The skin shows a brownish discoloration, and the con junctiva assumes a yellowish tint Associated with the disease is hypochromic anemia, leukopenia with a reduction of white cells to 4000 and below, and a decrease in platelets The peripheral lymph nodes are usually not swollen, but the internal nodes are frequently enlarged and show a brown discoloration

This disease must be differentiated from myelogenous leukemia, Hodgkin's and Banti's diseases It is not directly fatal, though the prognosis is poor, and the low vitality of the patient frequently leads to death from intercurrent diseases

Treatment Splenectomy has been suggested in cases of enlarged spleen and hemorrhagic diathesis with reduction of platelets No treatment is of any value so far as the involvement of the skeleton is concerned

REFERENCES

1 ALBRIGHT F DRAKE T G and SULKOWITCH H W *Bull Johns Hopkins Hosp* 51 158 1935

2 ASKENAZY, M *Arb a d Geb d path Anat und Bacteriol*, 4 398 1904
3 BARR D P BULGER H A and DIXON, H H *J A M A* 92 951 1929
4 BERGSTRAND H *Acta med Scandinav*, 76 128 1931
5 BOYD R T *Research Seminar Notes, Dept Orthop Surg*, State Univ of Iowa *14 D*, 1940-1941
6 CHRISTIAN H A *Contributions to Medical and Biological Research 1* 390, New York Hoeber 1919
7 EPSTEIN E *Biochem Ztschr* 145 398, 1924
8 ERDHEIM J *Frankfurt Ztschr f Path*, 7 168 1911
9 —— *Sitzungsber d k Akad d Wiss Wien Math Naturw Wissensch Klasse, 116* 311, 1907
10 —— *Rachitis und Epithelkörperchen* Wien 1914
11 FREUND E *Arch Surg*, 28 849 May 1934
12 GAUCHER D C E *De l Epithelioma Primitive de la Rate* These de Paris 1882
13 HAND A *Arch Pediat*, 10 673 1893
14 LETTERER E Frankfurt *Ztschr f Path* 30 377 1924 *Veroffentl a d Geb d Konstit Path* 8 114, 1934
15 LICHTENSTEIN L and JAFFE H *Arch Path* 33 777, 1941
16 —— *Arch Path* 30 993 1940
17 MANDELBAUM F S and DOWNEY H *Fol a haemat*, 20 139, 1916
18 MANDL F *Zentralbl f Chir* 53 260 1926
19 PAPPENHEIMER A M *J Exper Med* 64 965 1936
20 PICK L *Med Klin*, 18 1408 1922 *Am J Med Sc* 185 453 1933
21 RECKLINGHAUSEN F D *Festschr f R Virchow* 1891
22 ROWLAND R S *Arch Int Med* 42 611, 1928
23 SCHLUMBERGER H G *Mil Surgeon* 99 504 1946
24 SCHULLER A *Fortschr a d Geb d Roentgenstrahlen*, 23 2 1915
25 SIWE S *Ztschr f Kinderh* 55 212 1934
26 SNAPPER I *Med Clin Bone Dis* New York Interscience Publ 1943
27 TANNHAUSER S J *Lipoidosis* London Oxford Med Publ 1940
28 WEIL P *Zentralbl f Chir*, 49 1689, 1922

Lecture IV

ON AVASCULAR NECROSIS OR THE OSTEOCHONDRITIDES

IN CONTRAST to *inflammatory necrosis* which is always associated with a fibroblastic reaction and scar tissue formation, the *aseptic necrosis* results in a molecular absorption of dead bone and its gradual substitution by new bone. This process is called "schleichender Ersatz" by Axhausen,[1] or "creeping substitution" by Phemister.[24] A variety of causes may produce aseptic necrosis, such as traumatism following a fracture of the head of the femur, toxic agents as in caisson disease or in radiation necrosis, or circulatory or embolic factors as in the osteochondritis deformans coxae. Besides, interference with the synovial or the periosteal blood supply may cause a massive aseptic necrosis as, for instance, in joint fractures where fragments deprived of their blood supply succumb to necrosis and absorption. The fracture of the scaphoid very often results in necrosis of the proximal fragment, because its blood supply is precarious. Similarly, a fracture of the neck of the astragalus may so destroy the blood supply that the body of the bone succumbs to necrosis. It is generally assumed that bone transplants undergo aseptic necrosis, and that by a process of creeping substitution the graft is replaced by new formed bone from the recipient area.

The aseptic necrosis based on circulatory, embolic, or endocrine factors presents itself in a variety of clinical entities. There is the aseptic necrosis of the upper femoral epiphysis known as Perthes' disease, also the osteochondritis dissecans, an aseptic necrosis of the femoral condyle, first described by Koenig, Kohler's disease, an aseptic necrosis of the tarsal scaphoid, Haglund's disease, located in the calcaneal epiphysis, and many others. They all have in common the *mass necrosis* of a certain portion of bone, which is followed by a slow process of regeneration. The necrosis involves the cancellous tissues of the epiphysis, without destroying the general architecture of the bone. The radiographic appearance is not changed until the necrosis becomes manifest, by the contrast between its greater density and the lighter shadow of the surrounding atrophic bone.

Regeneration starts with active hyperemia in the neighboring bone, which sends capillary loops and fibrous stroma into the necrotic area. The dead bone is removed by osteoclasts carried by the invading bone marrow tongues. This is followed by the substituting and rebuilding of the haversian canals layer upon layer by the osteoblastic cells. It takes many months and sometimes years before this substitution is complete, and before the new formed bone regains its normal architectural pattern. It is significant for this type of necrosis that the cartilage which covers the epiphysis does not seem to suffer any degenerative changes, as it is adequately nourished by the synovial fluid.

On the other hand, during the process of substitution and regeneration the new bone is slow in calcifying, and the osteoid bone offers very little resistance to pressure stresses It is compressible, and under weight bearing or other stresses becomes deformed readily

The x ray diagnosis of the necrosis rests upon the increased density of the necrotic focus The beginning restitution is recognized by the appearance of areas of absorption and translucency, usually at points where the circulation enters into the necrotic area The penetration of the necrotic focus by marrow tongues with formation of osteoid bone also manifests itself by the so called fragmentation or the breaking up of the necrotic area in pieces, separated by translucencies Actual fractures within the necrotic bone occur under traumatic influences, and can also be seen in the x ray picture

I ASEPTIC NECROSIS OF THE HEAD OF THE FEMUR LEGG CALVE PERTHES' DISEASE, OR OSTEOCHONDRITIS DEFORMANS COXAE

This disease is known as Perthes',[23] Calve's,[6] or Legg's[19] disease, because it was first described by these three observers practically at the same time (1910) It is an aseptic necrosis of the head of the femur, which occurs in young individuals from six to fourteen years The necrosis is epiphyseal, and involves practically the entire head The necrotic head is demarcated from the living bone bv a hyperemic zone of fibrous tissue Within the necrotic head fracture lines appear

So long as the necrotic bone remains intact, it retains its thickness, its density, and its resistance against stress The penetration of marrow tongues into the necrotic area occurs both along the synovial vessels and through the center of the head, following the artery in the ligamentum teres As the osteoblasts are carried into the necrotic mass, bone is laid down around the necrotic lamellae, and the latter are absorbed by osteoclasts The new formed bone is osteoid and soft, and is very susceptible to deformation under static or mechanical stresses

A THE PATHOLOGY

The earliest changes occur in the soft tissue, the synovial membrane, the capsule, and the periosteum These tissues are swollen, edematous, and hyperemic, and the synovia becomes redundant at the inferior angle between head and neck

Microscopically one observes edema, hypervascularity, and thickening of the synovia, with villous formation There is perivascular lymphocytic and plasma cell infiltration Later the vascularity decreases, leaving a fibrous thickening of the soft tissues The mass of the head is necrotic, and in later stages one sees the substitution bv invading blood vessels and granulation tissue from the surrounding bone The head itself tends to become wider, and the neck

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IN CONTRAST to *inflammatory necrosis* which is always associated with a fibroblastic reaction and scar tissue formation, the *aseptic necrosis* results in a molecular absorption of dead bone and its gradual substitution by new bone This process is called "schleichender Ersatz" by Axhausen,[3] or "creeping substitution" by Phemister [4] A variety of causes may produce aseptic necrosis, such as traumatism following a fracture of the head of the femur, toxic agents as in caisson disease or in radiation necrosis, or circulatory or embolic factors as in the osteochondritis deformans coxae Besides, interference with the synovial or the periosteal blood supply may cause a massive aseptic necrosis as, for instance, in joint fractures where fragments deprived of their blood supply succumb to necrosis and absorption The fracture of the scaphoid very often results in necrosis of the proximal fragment, because its blood supply is precarious Similarly, a fracture of the neck of the astragalus may so destroy the blood supply that the body of the bone succumbs to necrosis It is generally assumed that bone transplants undergo aseptic necrosis, and that by a process of creeping substitution the graft is replaced by new formed bone from the recipient area

The aseptic necrosis based on circulatory, embolic, or endocrine factors presents itself in a variety of clinical entities There is the aseptic necrosis of the upper femoral epiphysis known as Perthes' disease, also the osteochondritis dissecans, an aseptic necrosis of the femoral condyle, first described by Koenig, Kohler's disease, an aseptic necrosis of the tarsal scaphoid, Haglund's disease, located in the calcaneal epiphysis, and many others They all have in common the *mass necrosis* of a certain portion of bone, which is followed by a slow process of regeneration The necrosis involves the cancellous tissues of the epiphysis, without destroying the general architecture of the bone The radiographic appearance is not changed until the necrosis becomes manifest, by the contrast between its greater density and the lighter shadow of the surrounding atrophic bone

Regeneration starts with active hyperemia in the neighboring bone, which sends capillary loops and fibrous stroma into the necrotic area The dead bone is removed by osteoclasts carried by the invading bone marrow tongues This is followed by the substituting and rebuilding of the haversian canals layer upon layer by the osteoblastic cells It takes many months and sometimes years before this substitution is complete and before the new formed bone regains its normal architectural pattern It is significant for this type of necrosis that the cartilage which covers the epiphysis does not seem to suffer any degenerative changes, as it is adequately nourished by the synovial fluid

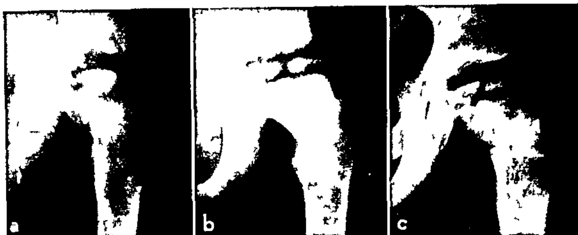


Fig 229 Perthes disease Case E L a) Necrosis b) Absorption c) Fragmentation and repair

recede from the acetabular cavity A more definite feature is a change in the epiphyseal line, which is decalcified at its junction with the neck, and has irregular contours Signs of necrosis do not appear until several weeks or even months after the onset The larger portion or sometimes the entire head appears as a dense, white necrotic mass (Figs 228a and 229a) Gradually within a matter of months, the decalcification of this necrotic head begins as the marrow granulations wander in and substitute new formed bone (Figs 228b and 229b) We see that during this substitution process the entire head becomes wider and thinner, especially if weight bearing has been permitted during this time At the same time, the neck appears thick and shortened, and its outer contour becomes convex The fragmentation of the head appears later, and is the sign of an already progressing reorganization (Figs 228c and 229c) Normal trabeculation in the new formed head appears very late If the head has been allowed to become deformed into mushroom shape, it remains in subluxated position Sometimes changes in the acetabulum can be seen as scalloped irregularities but, as a rule, the acetabulum follows the contours of the head It is only later that secondary changes of osteoarthritic nature appear

D THE TREATMENT

Osteochondritis deformans is a self limited disease which takes from two and one half to three and one half years, sometimes more, for complete resti



Fig 230 Osteochondritis deformans of the hip Case J J a) 1943 b) 1945 c) 1948

shorter and thicker Gradually the new formed bone becomes recalcified, though the head very often assumes a mushroom shape and no longer fits snugly into the acetabulum

B THE CLINICAL PATHOLOGY

1 General

It was believed at first that the osteochondritis deformans of the hip selects patients of a special type of anatomic build Froelich described such a type of obesity and hypogenitalism This idea that the disease occurs mostly in Froelich's type has now been abandoned, because many more patients are seen who are of perfectly normal anatomic build than of the so called Froelich syndrome

This condition is much more common in boys than in girls, the ratio being 85 15 (Howorth[11]) Unilateral cases also prevail over bilateral cases, but bilaterality is not uncommon

2 Local symptoms

The earliest symptom is the *limp*, which is at first intermittent, but gradually increases in intensity *Pain* also is not constant, but intermittent, especially in the beginning, only later does it become more persistent The pain is referred to the groin or the medial aspect of the thigh or knee, following the distribution of the obturator nerve It becomes worse with activity, and is definitely relieved by rest

Limitation of motion occurs early Abduction and internal rotation are restricted The joint is slightly tender anteriorly, and the Patrick sign is often positive One of the earliest symptoms is the Trendelenburg sign *Atrophy* of *the thigh is common Shortening occurs only later, and is always insignificant*

C THE X-RAY SIGNS

The x ray signs appear relatively late The first sign is produced by the reaction of the capsular apparatus, and consists in a bulging of the capsular sac Sometimes the joint space appears larger so that the head seems to



Fig 278 Perthes disease Case R D a) Massive necrosis b) Beginning substitution c) Fragmentation

II ASEPTIC NECROSIS OF THE TARSAL SCAPHOID KÖHLER'S DISEASE

This peculiar localization of aseptic necrosis was first described by Kohler [15] It is a rare condition and is found more often in younger individuals, some times in young children It is usually mistaken for tarsal tuberculosis The first signs are tenderness and localized swelling in the region of the tarsal scaphoid, associated with some restriction of motion The x ray picture shows the characteristic shadow of the necrotic scaphoid The bone is flat and thin,



Fig 232 (Left) Traumatic arthritis following osteochondritis deformans of the hip Case C S
Fig 233 (Right) Kohler s disease of the tarsal scaphoid Case J L

and its dense shadow contrasts strongly with the atrophy of the surrounding tarsal bones

This disease runs a definite cycle, and complete substitution always occurs within a period of one and one half to two years (Fig 233) The treatment consists in immobilization of the foot and elimination of weight bearing until regeneration is complete The patient usually recovers without any residual deformity or functional impairment

III ASEPTIC NECROSIS OF THE CARPAL SEMILUNAR KIENBOCK'S DISEASE

Almost at the same time Kienbock[14] and Preiser [5] described a condition involving the carpal lunate, which is similar to the above mentioned disease of the tarsal scaphoid It is essentially an aseptic necrosis There is some question whether the condition described by Preiser does not present a healing stage of a fissure fracture of this bone

However, in the case of Kienbock's disease, there is little doubt that it is a nontraumatic aseptic necrosis It usually involves younger or middle aged people, and is frequently found in persons who are working with compressed

tution (Figs 230 and 231) It is generally agreed that immobilization and elimination of weight bearing are essential Rest in bed and simple traction is the best means of preserving the head and avoiding secondary deformation, this treatment is advocated particularly by Waldenstrom It is most important in the incipient stage, where the signs are acute, and should be continued into the later stages when the decalcification of the head begins, and the structure of the head is soft and pliable In the acute stage traction not only relieves pain but also corrects the flexion contracture As long as



FIG 231 Perthes disease Case C P a) October 1932 b) May, 1934

pain and discomfort exist, recumbent treatment with traction should be continued

When the x ray picture shows the beginning regeneration of the head, it is permissible to change to ambulatory treatment, provided weight bearing can be eliminated This can be done in various ways The hip can be maintained in flexion position by a sling over the knee attached to a Sam Browne belt (Howorth[11]) It is safer, however, to resort to plaster cast and crutches, which prevent movement in the hip and also weight bearing, especially if the cast is applied with the knee in flexion

Under this conservative treatment our results in 94 cases, ranging in age from three to twelve years, treated between 1916 and 1936, and observed for a sufficient length of time were good in 74 per cent (Obletz[21]) In a series of 108 cases treated between 1935 and 1946 (Kaplan[13]) similar favorable results were obtained

To hasten the process of regeneration, some surgeons advocate drilling the head, similar to the procedure in the slipped epiphysis In view of the fact that the disease is self limited and that the reorganization of the head takes place spontaneously in due time, we doubt the wisdom of any operative measure The usual technique is to expose the hip by the Smith Peterson incision, then through a window made in the neck anteriorly, at the margin of the head, several drill holes are placed The advocates of this operative method claim that the end results are better, in comparison with conservatively treated cases The late after effects of the deformed head usually appear after many years in the form of traumatic arthritis (Fig 232)

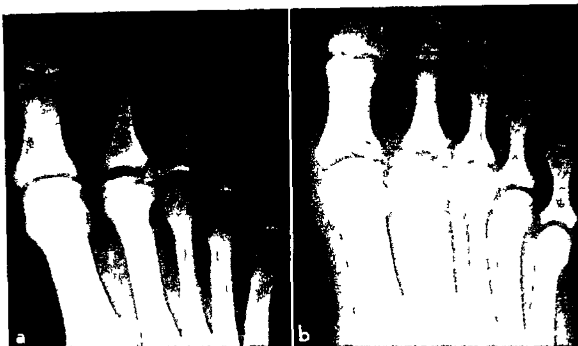


Fig 236 Kohler Freiberg's disease of the second metatarsal Removal of fragment of the head Case N H a) Before b) After

in the stage of substitution, and the fragmentation of the head suggests fracture In fact, in one case (Fig 236) we removed a fragment of the osteochondritic head because it interfered with free motion There was no history of trauma The shaft of the second metatarsal is usually considerably thickened, and appears more dense and sclerotic than the rest The entire neck seems to take part in the transformation

V OSGOOD SCHLATTER'S DISEASE

Here is another clinical entity of the osteochondritis group with a double name In the same year (1903) Schlatter [7] and Osgood described a condition of fragmentation and growth disturbance, involving the tibial tubercle



Fig 237 (Left) Osgood Schlatter's disease bilateral

Fig 238 (Right) Epiphysitis vertebrae Case O H

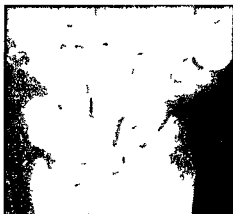


Fig 234 Kienbock s disease of the carpal lunate Case G H

air tools, the condition being ascribed to the continuous concussion of the bone The patient complains of pain and swelling and loss of strength in the hand The characteristic pressure point is at the dorsum of the wrist, midway between the radial and ulnar borders over the first carpal row, which corresponds to the situation of the lunate

In the x ray picture the bone appears already in a state of fragmentation, and the necrosis is demonstrated by the dense white shadow (Fig 234) The bone appears compressed Gradually the substitution of the bone sets in, although in cases in which the disease has followed acute trauma we have observed that the necrosis persists In the usual case, however, one may expect a gradual reorganization by creeping substitution in the course of one and one half to two years (Fig 235)

The treatment is immobilization in early cases Exceptionally, when the necrosis persists or when a secondary arthritis develops, it is necessary to remove the bone



Fig 235 Kienbock s disease a) 1945 b) 1947

IV OSTEOCHONDRITIS OF THE METATARSAL HEADS KOHLER FREIBERG'S DISEASE

This disease owes its hyphenated name also to almost simultaneous reports by Freiberg[9] and Kohler[10] in 1914 Here the seat of the aseptic necrosis is in the heads of the metatarsal bones At first it was believed to be an infection, but the subsequent course and cycle shows that it belongs to the group of aseptic necroses The usual complaint is tenderness and swelling over the anterior arch, more often in the region of the second metatarsal head

The x ray picture shows definite osteochondritic changes, often already



FIG 236 Kohler Freiberg's disease of the second metatarsal Removal of fragment of the head Case N H a) Before b) After

in the stage of *substitution, and the fragmentation of the head suggests fracture* In fact, in one case (Fig 236) we removed a fragment of the osteo chondritic head because it interfered with free motion There was no history of trauma *The shaft of the second metatarsal* is usually considerably thickened, and appears more dense and sclerotic than the rest The entire neck seems to take part in the transformation

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FIG 237 (Left) Osgood Schlatter's disease bilateral

FIG 238 (Right) Epiphysitis vertebrae Case O H

Schlatter considered it an epiphysitis of the descending tongue of the tibial epiphysis, rather than an avulsion Watson Jones believes that it is not a disease at all, but simply a traumatic condition comparable to any other epiphyseal separation However, it is certain that the condition undergoes a similar cycle of reconstruction as do the other types of osteochondritis The x ray picture usually shows the condition to be in a state of fragmentation (Fig 237) On microscopic examination one sees an area of hemorrhage between the separated portions in the tubercle This hemorrhage gradually becomes organized by the invasion of fibroblasts Cole[7] also describes an increased vascularity of the tendon at its attachment to the tubercle, as well as a roughening and irregularity at the line of attachment of the tendon to the tubercle

The x ray picture (Kriedelbaugh and Wyman[16]) is confusing, because normally the fusion of the apophysis with the shaft occurs rather late Nevertheless, an actual necrosis exists, whether of traumatic nature or not, and the substitution occurs by formation of new osteoid bone

Bosworth[3] distinguishes an acute and a chronic stage In the former the entire apophysis appears as a continuous but hazy mass In the chronic stage separate bony masses are seen Most likely several different conditions are included under the term Osgood Schlatter's disease as it is commonly applied In one instance there is a simple separation of the apophysis with fragmentation In another there is fragmentation and necrosis with or without epiphyseolysis In the true Osgood Schlatter's disease the necrosis heals by creeping substitution The patients are usually boys from ten to fourteen years They complain of pain in the region of the tibial tubercle, particularly on forced flexion of the knee or on kneeling Examination shows swelling and definite tenderness over the tibial tubercle The condition is sometimes bilateral

The treatment is simple and consists in immobilization in a plaster cast for two to three months By this time the symptoms have usually disappeared, and the spontaneous repair is well underway

VI OSTEOCHONDRITIDES OF THE SPINE

A VERTEBRAL EPIPHYSITIS SCHEUERMANN DELAHAYE'S DISEASE

Scheuermann's first description[26] (*1921*) antedates *that* of *Delahaye*[9] (1924) by three years The condition is an osteochondritis of the epiphyses of the vertebral bodies, and is also known as kyphosis dorsalis adolescentium or juvenilis

In involving the vertebral epiphysis, the disease contrasts with osteochondritis of the vertebral body proper The latter is a disturbance of the enchondral ossification within the body and appears much earlier when the ossification of the body is still in progress, usually between the third and fifth years, that is, during the second period of rapid growth It was first described by

Calvé,[5] and is known as osteochondritis vertebrae, to distinguish it from Scheuermann's disease, which is an osteochondritis of the vertebral epiphysis

The primary ossification centers of the body appear at the forty fifth day of pregnancy, and are all present by the fourth intra uterine month The secondary epiphyseal centers, however, do not appear until the eleventh and twelfth years of life, and do not fuse with the body until past maturity

In *vertebral epiphysitis* the vertebral epiphysis is fragmented, and ultimately unites with the body of the vertebra after some delay During this time the spine loses its resistance against weight bearing, the vertebral bodies become deformed particularly in the dorsal spine, and one sees frequent herniation of the intervertebral disc in the cancellous bone, which indicates that certain degenerative changes must occur in the disc also (Fig 238) The juvenile kyphosis usually involves young individuals from twelve to fourteen years, of rather slender and asthenic build The patient complains often of vague pain, and easy fatigue The pain is located along the spinous processes, and there is tenderness to pressure but no specific trigger point The dorsal and dorso lumbar portions of the spine are the ones most frequently affected (Fig 239) A dorsal or dorso lumbar kyphosis develops, and the lumbar spine is flattened Usually there is a corresponding sagging of the thorax, and the vital capacity is definitely diminished

In the course of time the pain disappears but the deformity increases and becomes rigid so that by the time the child is fifteen or sixteen years of age no further correction can be obtained

The treatment follows two purposes, first, the correction and, second, the relief of pain Correction is possible only in the earlier phases of the disease, so long as the spine still maintains some mobility It is carried out by recumbency or corrective casts Telson's method consists in the application of a cast in forward flexion, in order to abolish first any lumbar lordosis Then the dorsal spine is extended backward over the upper posterior edge of the plaster jacket This procedure seems adequate only in early cases In later stages one must be content with applying a spinal brace, which relieves the pain and prevents further deformation of the spine This should be accompanied by systematic development of the back muscles by means of physiotherapy and exercises (Figs 240 and 241)



Fig 239 Epiphysitis vertebrae Case B R

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B OSTEOCHONDRITIC VERTEBRA

In contrast to the epiphysitis, the osteochondritis involves the vertebral body and not the vertebral epiphysis It is much rarer than the epiphysitis The original description of Calve cites two cases Buchman[4] in an excellent review emphasizes that while the vertebral epiphysitis and the osteochondritis are parallel conditions, they appear in different periods of life, the osteochondritis appearing in the earlier period from two to six years The symptoms are moderate, there is some pain, tenderness, spasm, and limitation of motion The disturbing factor is the deformity of the back which may appear in the form of a knuckle or an arc, depending on the number of vertebrae involved

The x ray picture reveals an irregularity of the vertebral outlines and a flattening and wedging of the vertebrae This is then followed by a stage of restitution in which the vertebral outlines again appear dense and sclerosed, but *the deformation of the vertebra is final* Corresponding with the thinning of the vertebral bodies, one finds the intervertebral spaces rather enlarged and widened

The osteochondritis vertebrae as well as the vertebral epiphysitis develops on the basis of the intrinsic changes in the vertebral bodies or epiphyses respectively They have nothing to do directly with muscular or ligamentous insufficiencies However, once in progress, these conditions may be greatly accentuated by external causes, such as strain or exertion

So far as the treatment of this condition is concerned, recumbency is indicated in earlier stages, and a plaster jacket afterward, when the painful stage has passed *If a deformity remains* as is usually the case, it does not interfere with the activity or stability of the spine

VII HAGLUND'S DISEASE OR OSTEOCHONDRITIS OF THE CALCANEAL APOPHYSIS

Haglund[16] described an osteochondritic lesion which occurs in the apophysis of the os calcis The local symptoms are slight swelling and tenderness The x ray picture (Fig 242) shows in the earlier stages a dense shadow suggesting aseptic necrosis Later a definite fragmentation of the calcaneal apophysis is seen This disease is easily controlled by immobilization



Fig 242 Haglund's disease in a case of osteopoikilosis

A similar condition affecting the patella has been described also by Larsen[17] and Johansson[1]

Fig 240 Epiphysitis vertebrae Case J D a) January 1947 b) October 1947

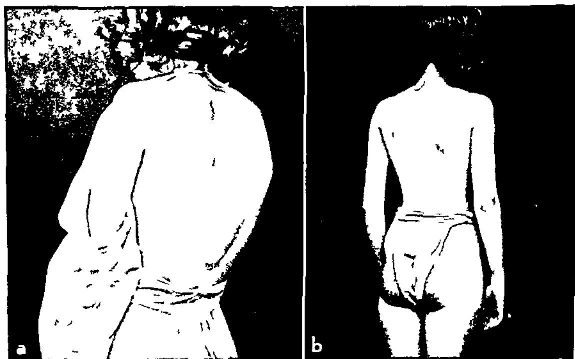


Fig 241 Epiphysitis vertebrae Case D E a) April 1939 b) June 1941

Lecture V

ON NEURO-ARTHROPATHIES

I DEFINITION

SINCE Charcot[1] described in 1868 the arthropathic joint in tabetics, it has become commonly identified with this disease As a matter of fact, the arthro pathic or Charcot joint is encountered in many other conditions It was observed in hemiplegia bv Scott and Allison even as early as 1843, and in trans verse myelitis by Magnier in 1859 Packard found (1863) that lesions of peripheral nerves produce "arthropathic" joints, in the same year Weir Mitchell reported similar joint changes in lesions of the spinal cord Later Schlesinger demonstrated the occurrence of arthropathic joints in syringo myelia

II THE ETIOLOGY

Is the arthropathic joint a trophic disease entity, or does it represent the effect of traumatism on a joint with impaired sensibility? It was Charcot's idea that the disease was of purely trophic character due to the change in the posterior roots and the cord seen in tabes In opposition to this view it can be argued that there are no such trophic degenerative changes in syringomyelia, where loss of sensibility and vasomotor changes obviously suffice to produce a Charcot joint In this respect we refer to the experiments of Eloesser, who found that joint changes corresponding to the Charcot joint could not be produced simply by cutting the posterior roots and causing anes thesia, but that they did develop when the analgesic joint was traumatized He concluded that it is the trauma in combination with the joint anesthesia which is responsible for the development of the neuropathic joint This is significant from the prophylactic and therapeautic point of view, because it shows how important it is to protect such joints from strain and injury

While trauma is never the cause, yet it is undoubtedly an essential con tributing element in the course of neuropathic joints The point is, that because of the insensibility of the joint, little attention is paid to trauma, and there is no warning sensation of pain to protect the patient against harmful external forces What may seem to him a slight trauma or strain often pro duces a massive swelling with tension of the skin but with little disturbance of function After a presumably slight twist the patient may find himself with a fractured bone but still able to walk In arthropathic joints trauma must be evaluated differently from other situations One must realize that fracture of bones or collapse of joints occurs without extraordinary hazard In fact, pathological fractures may be the earliest clinical sign of tabes

VIII OSTEOCHONDRITIS OF THE ELBOW JOINT PANNER'S DISEASE

The site of aseptic necrosis is occasionally the capitellum of the humerus (March[19]) It was first described by Panner, and is known as Panner's disease It occours very rarely A similar condition, an osteochondritis of the supra trochlear septum, is described by Morton and Crysler,[20] who reported six cases It is characterized by recurrent attacks of pain and tenderness above the tip of the olecranon or the upper part of the antecubital space, and is associated with limitation of motion

The x ray shows a sharply demarcated circular fragment of bone, located in the supratrochlear fossa Microscopically necrotic foci can be seen underneath the hyaline cartilage of the trochlea

REFERENCES

1 AXHAUSEN, G *Deutsche med Wchnschr* *39* 111 1914
2 ——— *Virchows Arch f path Anat* *252* 458 1924
3 BOSWORTH C M *Am J Surg* *43* 526 1939
4 BUCHMAN J *J Bone & Joint Surg* *9* 55 1927
5 CALVE J *J Bone & Joint Surg* *7* 41 1925
6 ——— *Rev de Chir* *42* 54 1910
7 COLE J P *Surg Gynec & Obst* *65* 55 1937
8 DELAHAYE A These de Paris 1924 *Presse med* *72* 137 1924
9 FREIBERG A H *Surg Gynec & Obst* *19* 191 1914
10 HAGLUND T *Arch f klin Chir* *82* 922 1907
11 HOWORTH M B *J Bone & Joint Surg* *30 A* 601 1948
12 JOHANSSON S *Hygiea* *34* 161 1922
13 KAPLAN N *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *18 D* 25 1947
14 KIENBOCK R *Fortschr a d Geb d Roentgenstrahlen* *16* 77 1910
15 KOHLER A *Munchen med Wchnschr*, *372* 1922 1908 *Fortschr a d Geb d Roentgenstrahlen* *22* 512 1914 1915
16 KRIEDELBAUGH W W and WYMAN A C *Am J Surg* *75* 553 1948
17 LARSEN *Norsk mag f laegevidensk* Publ in *Nord Med* *82* 856 1921
18 LEGG A T *Boston M & S J* *162* 202 1910
19 MARCH H C *Am J Roentgenol* *51* 682 1944
20 MORTON H S and CRYSLER W E *J Bone & Joint Surg* *27* 12 1945
21 OBLETZ B E *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *10 D* 44 1936
22 OSGOOD R B *Boston M & S J* *148* 114 1903
23 PERTHES G C *Deutsche Zt Chir* *107* 111 1910
24 PHEMISTER D B *J Bone & Joint Surg* *12* 769 1930
25 PREISER G *Fortschr a d Geb d Roentgenstrahlen* *17* 360 1911
26 SCHEUERMANN H B *Ztschr f orthop Chir* *41* 305 1921
27 SCHLATTER C *Beitr z klin Chir* *38* 874 1903

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REFERENCES

1 Axhausen G *Deutsche med Wchnschr* *39* 111 1914
2 ——— *Virchows Arch f path Anat* *252* 458 1924
3 Bosworth C M *Am J Surg* *45* 526 1939
4 Buchman J *J Bone & Joint Surg* *9* 55 1927
5 Calvé J *J Bone & Joint Surg* *7* 41 1925
6 ——— *Rev de Chir* *42* 54 1910
7 Cole J P *Surg Gynec & Obst* *65* 55 1937
8 Delahaye A. These de Paris 1924 *Presse med* *72* 737 1924
9 Freiberg A H *Surg Gynec & Obst* *19* 191 1914
10 Haglund T *Arch f klin Chir* *82* 922 1907
11 Howorth M B *J Bone & Joint Surg* *30 A* 601 1948
12 Johansson S *Hygiea* *34* 161 1922
13 Kaplan N *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *18 D* 25 1947
14 Kienbock R *Fortschr a d Geb d Roentgenstrahlen* *16* 77 1910
15 Kohler A *München med Wchnschr* *372* 1922 1908 *Fortschr a d Geb d Roentgenstrahlen* *22* 512 1914 1915
16 Kriedelbaugh W W and Wyman A C *Am J Surg* *75* 555 1948
17 Larsen *Norsk mag f laegevidensk* Publ in *Nord Med* *82* 856 1921
18 Legg A T *Boston M & S J* *162* 202 1910
19 March H C *Am J Roentgenol* *51* 682 1944
20 Morton H S and Crysler W E *J Bone & Joint Surg* *27* 12 1945
21 Obletz B F *Research Seminar Notes* Dept Orthop Surg State Univ of Iowa *10-D* 44 1936
22 Osgood R B *Boston M & S J* *148* 114 1903
23 Perthes G C *Deutsche Zt Chir* *107* 111 1910
24 Phemister D B *J Bone & Joint Surg* *12* 769 1930
25 Preiser G *Fortschr a d Geb d Roentgenstrahlen* *17* 360 1911
26 Scheuermann H B *Ztschr f orthop Chir* *41* 305 1921
27 Schlatter C *Beitr z klin Chir* *38* 874 1903

forcing capsular ligaments, such as the collateral ligaments of the knee, as well as destruction of the intra articular ligamentous structures, such as the menisci, the cruciates in the knee, or the ligamentum teres in the hip joint

The *para articular changes* appear in form of massive osteophytes, bridges, and buttresses of bone irregular in shape, as seen for instance at the condyles of the femur and around the acetabulum, they often form the so called ring ankyloses or ankyloses cerclees around the joint

Juxta articular bone formations are the ossifications in the soft tissues outside the joint They form large, trabeculated masses of bone devoid of any static orientation These bone masses are completely encapsulated in fibrous tissue which separates them from their muscle bed

The *peripheral nerves* show interstitial and parenchymatous neuritis and degeneration with crumbling of the myelin and disappearance of the axis cylinders



Fig 245 Pannus adherent to cartilage in a nonpressure area

B MICROSCOPIC PATHOLOGY

Virchow called attention to the analogy between arthropathy and hyper trophic arthritis In both conditions an essential feature is the revival of the enchondral ossification at the cartilage bone border

1 The first changes are noticed in the *cartilage* The superficial horizontal strata are increased in thickness, and the cells are grouped in Weichselbaum's lacunae (Fig 243) The provisory calcification zone is broadened Later, there appears a coarse, perpendicular fibrillation with loss of the gliding layer, the zone of provisory calcification begins to show perforation by marrow tongues (Fig 244)

2 In areas of the joint not exposed to pressure a *pannus* makes its appear ance, arising from the synovial membrane It is firmly adherent to cartilage which becomes dissolved beneath it (Fig 245)

3 In *pressure areas* the subchondral spongiosa becomes absorbed, and the

III THE PATHOLOGY

A GROSS PATHOLOGY

The outstanding feature of the arthropathic joint is the bizarre combination of *productive bone changes* within the joint and the capsular apparatus and a *rapid* and *massive destruction* of bone Destructive changes include massive necrosis of the joint constituents, fragmentation of the articular surfaces, formation of free bodies, degeneration and destruction of cartilage with erosions, and destruction and absorption of large masses of bone adjacent to the articular surfaces On the other hand, the productive changes consist in massive apposition of bone around the joint periphery and in the capsular and pericapsular tissues

The *joint capsule* shows degeneration of the synovial membrane, fibrous infiltration of the capsular wall, and relaxation and destruction of the rein



FIGS 243 and 244 Tabetic joints cartilaginous changes Figure 243 shows broadening of the provisory calcification zone and grouping of the cartilaginous cells in Weichselbaum s lacunae the superficial or horizontal stratum of the joint cartilage is increased in thickness Figure 244 shows fibrillation of the degenerated joint cartilage and loss of the gliding layer perforation of marrow tongues through the zone of provisory calcification



FIG 248 Tabetic joint Staggering ossification line, central exostosis In places the joint cartilage disappears completely and is substituted by bone

(Fig 249), or they come from the periosteum of the joint constituents under the influence of ligamentous or tendinous tension (Fig 250)

6 *The effect of trauma Wear and tear* causes fraying of the pannus and fissuring of the cartilage Pressure produces a thinning of the cartilage and a reactive thickening of the provisory calcification zone, and results in attrition of the subchondral bone *Joint fractures* cause depression and collapse of joint surfaces such as, for instance, the plateau fractures of the tibia The cartilage covering of fractured portions, however, continues its reactivated enchondral ossification

7 *The joint capsule* The fibrous capsule is thickened, it is very vascular in its deepest layers, and the lymph spaces are greatly increased The layers include masses of cartilage and bone (Fig 251), which continue to grow



FIG 249 (Left) Tabetic joint A marginal exostosis projecting in between the two joint bodies
FIG 250 (Right) Tabetic joint A marginal exostosis formed at the insertion of a ligament

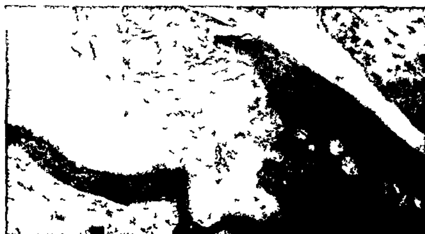


Fig 246 Joint cartilage and subchondral bone destroyed and substituted by connective tissue

marrow spaces are filled with connective tissue, owing to *fibrous degeneration of the bone marrow* (Fig 246)

4 The revival of the *enchondral ossification* manifests itself by the perforation of the provisory calcification zone by numerous marrow tongues They dissolve the matrix of the cartilage zone and lay down enchondral bone (Fig 247) Not only all the hyaline cartilage is finally used up in the process of enchondral ossification, but the fibrocartilage produced by the pannus also undergoes a similar transformation

5 This ossification process leads to formation of pedunculated *central exostoses* which project into the joint They are flat and mushroom like (Fig 248) *Marginal exostoses* form from the marginal portions of the cartilage

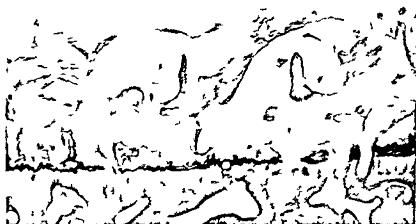


Fig 247 Tabetic joint Perforation of provisory calcification zone and ossification of the deep portion of the joint cartilage

B THE AGE, ONSET AND LOCALIZATION

The *ages* varied from 17 to 82 When first seen 85 per cent of the cases were between 40 and 70 The male sex prevailed, with 80 per cent

The *time interval* between the syphilitic infection and the appearance of the Charcot joint varied between five and 40 years The time between the appearance of tabes and the tabetic joint averaged two to three years

An *acute onset* of the tabetic joint symptoms following fracture was observed in 28 cases or 20 90 per cent, following other injuries, in 31 cases or 23 38 per cent, thus, in a total of 59 cases or 44 27 per cent a precipitating traumatic factor could be established In all other cases the onset was gradual and spontaneous

In only 23 out of 134 cases, or 17 per cent, was a *symmetrical joint* involvement noted, although it is assumed that bilateral lesions occur with a frequency of one in four cases At any rate, symmetrical involvement is more frequent in tabes than in syringomyelia This may be explained by the symmetrical lesions of the posterior spinal tracts in tabes, in contrast to the more asymmetrical destruction of gray matter in syringomyelia

C THE LOCAL SIGNS

It is erroneous to believe that the neuropathic joint is a late occurrence, following the general signs of tabes after a long interval On the contrary, most tabetic arthropathies and especially the spontaneous fractures occur in the pre ataxic stage The same is true of syringomyelia, in which according to Schlesinger arthropathies often are an early symptom, preceding other symptoms by many years

In no less than 76 of our series of 134 cases, or 57 per cent, the joint lesion was the first complaint of the patient Of these 76 joints, 43 developed after fracture or some other trauma

1 Effusion and swelling is most frequently observed as an early sign It is painless and without redness, and often disappears leaving only a periarticular thickening A common cause of sudden effusion is a joint fracture, although effusion without fracture is not infrequent, especially in the more benign cases According to Oehlecker, these are cases of essentially synovial type We observed early effusions in 102, or 76 per cent, of the cases

2 Tenderness and pain While the swelling is, as a rule, associated with analgesia of the joint, the synovial tissue still retains some tenderness, and complete analgesia exists only in cases of longstanding Consequently, *pain on weight bearing* is not unusual We observed it in 48 cases, or 35 per cent, and as an early symptom in 14, or 10 4 per cent

3 Abnormal mobility is, next to swelling and effusion, the most important local symptom It may proceed to the point of complete flailness The principal cause is the progressive destruction of the joint Sometimes the flailness develops very rapidly, within a few months, though the patient is still able to

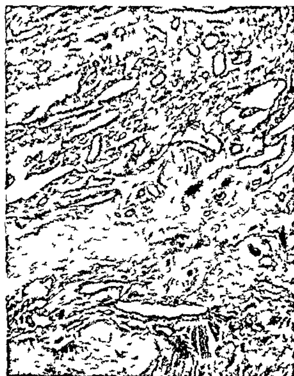




Fig 252 Tabetic joint A pedicled body of bone coming from the inner surface of the joint capsule



Fig 251 Tabetic joint capsule greatly thickened very vascular in its deeper portions (mostly lymph spaces) Peripheral portions are fibrotic

within the fibrous tissue of the capsule Pedicled bodies of bone formed in the capsule may become separated and form free bodies in the joint (Fig 252)

IV THE CLINICAL PATHOLOGY

According to Lotheisen 10 per cent, and to Marie 4 to 5 per cent of all tabetics acquire arthropathic joints, Schlesinger estimates the frequency in syringomyelia as 25 per cent In tabes the lower extremities are most often involved, the sequence is knee, ankle, and hip On the other hand, among 150 syringomyelitic arthropathies of Schlesinger, 122 involved the upper extremities, in sequence, the shoulder, elbow, hand, and fingers

A THE FREQUENCY OF GENERAL TABETIC SYMPTOMS IN ARTHROPATHIES

We examined 134 cases totalling 214 Charcot joints, all of which with the exception of two were due to tabes The *Wassermann reaction* (106 observations) was positive in 61 32 per cent, and negative in 38 67 per cent, of the latter group, however, the Kahn or Kline was positive in four cases and the synovial fluid Wassermann in 1 case We found a positive *Rhomberg sign* (62 observations) in 82 25 per cent and a negative Rhomberg in 17 74 per cent The *Argyle Robertson pupillary sign* (99 observations) was present in 80 per cent, and absent in 19 19 per cent

Among 112 cases the *knee reflexes* were absent in 82 14 per cent, and present in 17 86 per cent Only in nine cases or 7 3 per cent did the patients complain of definite *paresthesia and numbness* of the limbs *Lancinating pain* irrespective of pain about the joint was observed in 30 cases, or 22 per cent

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3 Abnormal mobility is, next to swelling and effusion, the most important local symptom. It may proceed to the point of complete flailness. The principal cause is the progressive destruction of the joint. Sometimes the flailness develops very rapidly, within a few months, though the patient is still able to

bear weight In all our cases of flail tabetic joints the process of bony destruction was found to be far advanced In 2 cases the tibia could be dislocated voluntarily, in a 3rd, the knee could be hyperextended to 45 degrees, and the hip of the same side was telescoping

4 Pathological fractures Spontaneous fractures without any external *violence are frequent in patients suffering from tabes Abnormal fragility of* bone is found in a number of other conditions affecting the central nervous system, in progressive paralysis, hemiplegia, spina bifida, and other congenital lesions of the cord, also in paraplegias from transverse cord lesions

These fractures heal usually as normal fractures would under sufficient immobilization, although functionally the callus may not be as sound as normal callus Also, because of analgesia and ataxia, undue strain is imposed prematurely upon the callus, which is often of the luxuriant type On the whole, a longer immobilization is necessary It is believed that these fractures occur most frequently in the stage of incoordination, but they may appear in earlier stages

Our 33 cases of pathological fracture constitute 24 8 per cent of the series They include fracture of the astragalus, os calcis, neck or shaft of the femur, ankle, condyles of the tibia or femur, humerus, and pelvis

5 Deformity in Charcot joints Considering the great amount of destruction in the joint, it is not surprising to see deformities of all kinds and degrees In 109 cases in which the knee joint was involved the most frequent deformity was genu varum, in 19 cases or 17 4 per cent, genu valgum was found in 14 cases or 13 per cent, genu recurvatum, in six cases or 5 8 per cent In 38 cases of ankle joint involvement varus deformity was observed in four cases or 10 5 per cent, and valgus in two cases or 5 2 per cent In the Charcot spine kyphosis and scoliosis were observed in *all 14* cases Shortening due to joint destruction ranged up to three inches In one instance of a Charcot hip it was due to absorption of the head and medial portion of the neck of the femur, in another a two inch shortening was caused by absorption of the head of the femur alone

6 Suppuration in Charcot joints Pyogenic infection superimposed upon a Charcot joint is a very serious complication It is usually not controllable Suppuration may follow infection from a perforating ulcer, as in the foot More often it is the result of direct ulceration of the skin over the joint, only rarely does it occur secondarily through the blood stream

The prognosis of suppurative arthritis in Charcot joints is usually poor The pyogenic infection controls the picture Ten of our cases became infected through ulcers and local sores All except two came to amputation Of the two cases in which the infection could be controlled, one involved the knee joint and control was accomplished by arthrotomy The other case was a neuropathic subastragalar joint in which the infection was controlled by wide drainage under Orr's technique

D THE X-RAY FINDINGS IN CHARCOT JOINTS

Generally speaking, the x ray signs consist in sclerosis, fragmentation, and absorption, in new bone formation and exostoses, in extra articular ossification, pathological fractures, dislocation, disalignment, and joint effusion All these phenomena have a certain pathological relationship, but it is not easy to recognize their sequence and significance in the course of this remarkably erratic and unpredictable affliction

1 Sclerosis

It is one of the early and most significant signs It always precedes and keeps ahead of the process of fragmentation and destruction In the knee, for

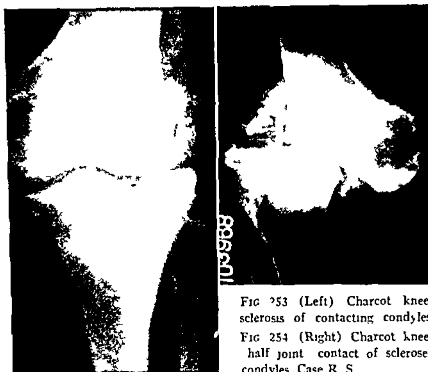


Fig 253 (Left) Charcot knee sclerosis of contacting condyles
Fig 254 (Right) Charcot knee half joint contact of sclerosed condyles Case R S

instance, when the articular cartilage is already completely disintegrated, the sclerotic process still extends to involve the femoral and tibial condyles in their entire width Where the process involves one half of the joint only, the contiguous medial or lateral condyles of the tibia and femur are involved in the sclerotic process (half joint contact, Figs 253 and 254)

Is the sclerosis reversible? It is possible for the sclerosis to disappear and to be substituted by more or less normal trabeculation and bone of normal density, but this occurrence is very rare We observed in one case that the sclerosis which involved the acetabulum cleared up after the head had succumbed to destruction (Fig 255)

The sclerosis is not uniform, it depends on static factors In the hip joint the greater portion of the acetabular region shows sclerosis, but the greatest density corresponds to the areas of support of the femoral head (Fig 256) As the latter wanders upward and laterally with the increasing destruction of



Fig 255 (Left) Charcot hip sclerosis of acetabulum that cleared up

Fig 256 (Right) Charcot hip melting of femoral head and sclerosis of acetabulum Case V F

the acetabulum, we see that the maximum of sclerosis wanders laterally also (Fig 257) If a supporting ledge or buttress is projecting from the acetabulum, the greatest amount of sclerosis is seen in this ledge or shelf

2 The atrophy and osteoporosis

Sclerosis which plays such an important part is associated not only with absorption, but also with regional atrophy and osteoporosis It must be understood that the sclerosis is strictly regional, and involves only the articular constituents Beyond these limits true atrophy can be seen, though it is never very marked We find it, for example, in the supracondylar regions of the femur As a rule, this atrophy is found only where there are also trophic changes of the overlying soft parts



Fig 257 Charcot hip wandering sclerosis of acetabulum Case G T

3 The disalignments and pathological dislocations

Can the ligamentous and capsular relaxation, caused by effusion of the joint, produce pathological dislocation or is it necessary that the joint be destroyed? We have observed only 1 case where the joint bodies were still intact and the contours

were apparently still normal, where nevertheless an anterior dislocation of the tibia (marked genu recurvatum) existed following a long period of joint effusion (Fig 258)

V SPECIAL CLINICAL SITUATIONS

A SKULL AND HEAD

On rare occasions one observes atrophy and spontaneous necrosis of the mandibles and the maxillae in tabes and in syringomyelia Rosin (1891) reported 22 cases in the literature There may be a loss of teeth without inflammatory signs, an analgesia of the gums, and extensive necrosis, but there is never any complicating phlegmon Anesthesia of the trigeminus is common, involvement of the motor root is rare Ulceration of the mouth occurs, and spontaneous fracture of the mandible has been seen, also pseudoarthrosis



Fig 258 Charcot knee subluxation but no gross joint destruction (X ray of case in Figure 267)



Fig 259 Charcot shoulder

Syringomyelitic lesions of the skull were reported by Schlesinger, as early as 1895

B TABETIC JOINTS OF THE UPPER EXTREMITY

The joints of the upper extremity are involved in 80 per cent of the syringo myelitic arthropathies, in contrast, only a small percentage of upper extremity arthropathies is encountered in tabes In our series of 134 cases involving 214 arthropathic joints, there were only five cases or 2 4 per cent involving the upper extremity, three elbows and two shoulders

In general, the *shoulder* is more frequently involved Extensive destruction of the humeral head is common, and is often followed by spontaneous dis location With the melting away of the head, massive effusion appears in the joint Marginal exostoses and para articular calcium deposits are seen (Fig 259) Erosion of branches of the axillary artery was observed in 1 case in our clinic (Milgram)

The *elbow* likewise shows severe changes There is an enlargement of the coronoid fossa, and the grinding off of the joint surfaces finally leads to complete destruction of the joint, resulting in a flail joint with pathological dislocation The articular and peri articular structures are enormously thickened Ample new bone formation and calcification occurs in the muscles, particularly in the triceps and the brachialis anticus Free bodies are frequently found (Fig 260) As many as 100 of these bodies have been observed, so that the picture resembles osteochondromatosis

There is no effusion in the *wrist and hand*, but there are, nevertheless, very marked changes in the distal end of the forearm bones and the carpals A



Fig 260 Charcot elbow



Fig 261 Charcot hip

volar dislocation of a single carpal bone or of the entire carpus may result The joint does not become flail, as the shoulder or elbow, but remains rather rigid Numerous exostoses can be seen The interphalangeal and metacarpophalangeal joints are occasionally affected The thumb, especially, shows arthropathic changes of the metacarpophalangeal joint

C TABETIC JOINTS OF THE LOWER EXTREMITY

1 The hip joint The *hip* is not as frequently involved as the knee or ankle, there were only 33 cases, or 13 per cent, in our series Flatow reports 38 cases among 149 tabetic arthropathies In syringomyelia, arthropathy of the hip joint is likewise rare, Schlesinger reports only one case in his series of syringomyelic arthropathies

The characteristic feature of the arthropathy of the hip is sclerosis of the acetabulum with bone proliferation and periarticular bone apposition, and a rapid melting down of the head (Fig 261) By an alternation of destruction and sclerosis, the acetabulum becomes enlarged (wandering acetabulum) The head follows, and the femur telescopes (Fig 262) While in some cases a sort of ankylosis may develop, the usual event is a complete melting down of the

head and neck, leaving only a conical end of the shaft (Charcot's so called baguette de tambour) Occasionally there are free bodies in the hip joint, more common is extensive ossification of the muscles, especially in the adductor group Posterior dislocation occurs after erosion and destruction of the posterior acetabular margin

2 The arthropathy of the pelvis is characterized by hyperplasia, and ossification of the ligments and of the muscle insertions, and formation of massive exostoses at the iliac bones and at the symphysis associated with destructive lesions of the pelvic joints In the sacro iliac joint the destruction may result in diastasis and complete disintegration

3 The knee Of all joints, the knee is most frequently involved in tabetic arthropathies In 109 of 214 tabetic joints, or 50 9 per cent, there was involvement of the knees The characteristic features of the arthropathic knee are the massive effusion and capsular relaxation, and the erosion and destruction of the joint bodies with marked deformities and disalignments In one type of case the disease develops slowly, simulating deforming arthritis, in another type the onset is sudden, with massive effusion and rapid disintegration The first type may cause considerable diagnostic difficulty, but the absence of pain or heat distinguishes it from a chronic arthritis



Fig 262 Charcot hip, telescoping femur Case F L



Fig 263 Charcot knee



Fig 264 Charcot knee

The arthropathies with sudden onset are usually the result of a preceding intra articular fracture, such as a chip fracture of the condyles The group with slow development may show secondary fractures later in the course

Early x ray signs are bone infraction or avulsion, slight contour changes of the condyles, exostoses, and absorption Later signs are depression of the condyles with formation of genu valgum and varum, proceeding to complete melting down of the condyles and flailness of the joint The end result is de



Fig 265 Tabetic genu varum



Fig 266 Charcot knee, genu valgum Case W S

struction of the joint, sometimes with posterior or lateral dislocation of the tibia (Figs 263 and 264)

Genu varum is the most common deformity (Fig 265) Of 39 cases the tabetic deformity of the knee was a genu varum in 19, a genu valgum in 14 (Fig 266), and a genu recurvatum in six (Fig 267)

4 The ankle was the joint involved in 38 of our series, or 17 7 per cent The extensive destruction and disintegration of the joint is associated with the formation of para articular exostoses Swelling, absence of pain, and de formity are the principal signs A pes varus is the most common deformity (Fig 268) Sometimes a malleolar fracture precedes the arthropathic changes (Fig 269) The combination of Charcot knee and ankle is frequent (Fig 270)

5 The tarsus In 15 cases, or 7 per cent the tarsal joints were affected Pathological shear fractures of the astragalus are observed Similar fractures



FIG 267 Tabetic genu recurvatum



FIG 268 Charcot ankle varus deformity Case I C

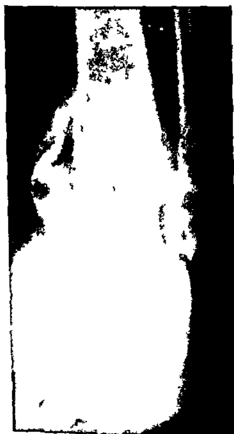


FIG 269 Charcot ankle following malleolar fracture



FIG 270 Bilateral Charcot knee and ankle



FIG 271 Charcot tarsus collapse of scaphoid and cuneiforms

of the scaphoid occur frequently In advanced cases all tarsal bones collapse (Fig 271) Depression and flattening often follows when the os calcis is involved (Fig 272) Less frequent are fractures of the metatarsals The so called *pie tabetique* is a shortened, thickened, and rounded foot in which the normal contours and curves are gone, and the arch of the sole is flattened

In the toes one finds the *metatarsophalangeal articulation* more often involved than the mid and end phalangeal joints (Fig 273) A *mal perforant,* described as a new disease by Nelaton (1872), is almost always present in advanced cases of tabes of the foot It is situated most often over the big toe and heel It takes its start from a neighboring neuropathic joint, the immediate cause being the pressure ischemia of the weight bearing areas of the foot



FIG ?7? Charcot tarsus collapse of os calcis

D THE CHARCOT SPINE

Spinal involvement is fourth in frequency among tabetic lesions of the skeleton The first two cases were described by Charcot (1886) Our series includes 14 cases, or 10 4 per cent of the total The preferred site is the lumbar and lower dorsal spine *The initial changes are sclerosis and formation of long spurs and exostoses*



Fig 273 Charcot metatarsophalangeal joint Case H H



Fig 274 Charcot spine earlier changes





Fig 275 (Left) Charcot spine later stage
Fig 276 (Right) Charcot spine (transverse myelitis) collapse and massive paravertebral bone formation

(Fig 274) This is followed by wedge formation and scoliotic deformity (Fig 275), and finally by total collapse and massive paravertebral bone apposition (Fig 276)

Compression of the cord and cauda equina are rare In syringomyelia one frequently sees scoliotic deformities in the cervicodorsal region Lumbodorsal scoliosis is observed in tabes

VI THE TREATMENT OF NEUROPATHIC JOINTS

A CONSERVATIVE

Most observers believe that the treatment should always be conservative, the principal measure being immobilization in casts or braces, and elimination of all joint motion No spontaneous healing is possible, but the arrest of tabetic destruction is often accomplished by early application of supportive apparatus This applies especially to the knee joint

For immobilizing the *knee joint* we recommend a well fitting long leg brace with a tuber seat arrangement Fifty six Charcot knees were treated in this manner Even in bilateral cases long leg braces are well tolerated, and the patient is able to walk by means of two canes For the *ankle* we advise a short leg brace with a free ankle joint to be attached to the shoe For the *hip joint* a long leg brace with a tuber seat attached to a broad pelvic girdle is very useful, as it eliminates all weight bearing on the joint Twenty of our hip cases were treated by these conservative procedures The *tabetic spine* requires immobilization in a spinal brace of the Taylor type or, in advanced cases, the more rigid and substantial standard body brace with a rigid anterior and posterior frame For the *elbow* a long arm brace supplied with a hinge joint of limited range of motion offers sufficient protection

B OPERATIVE

1 Fusion and resection

Conservative treatment means only the protection of the afflicted joint and the retardation, and sometimes the arrest, of the disease Can the disease be eliminated by resecting the joint? Ullman, who first advocated the operative resection (1897), did not expect consolidation, and most of the earlier surgeons declined the operation However, more recently good results from the resection of tabetic joints were reported (Oehlecker)

It is necessary to select the cases with the greatest care, the most important point is the early diagnosis Only in earliest stages is resection of any avail It must be done before greater masses of bone have become destroyed, and while an accurate adaptation of broad, comparatively healthy bony surfaces is still possible One should not overlook that the tabetic joint is eminently susceptible to pyogenic infection, and that it is very poorly equipped to control it Practically all infected cases lead ultimately to amputation

For the hip we use the extra articular fusion, and in some cases, the Lorenz

osteotomy. The operation of choice for the knee is fusion rather than resection, but it is applicable in the very early cases only. More advanced cases must be resected. The bone ends should be freshened only sparingly, to simplify the operation we refrain from removing the joint capsule.

2. Amputation

There always remains a number of cases for which the amputation is the only solution. These are, first, the cases with suppurative arthritis of the knee joint. In our series there were seven amputations for Charcot knees, five because of suppurative arthritis. The other two were performed because of advanced destruction of the joint.

Another case for amputation is the Charcot ankle with massive destruction, which can no longer be controlled by supportive apparatus. In this case Syme's supramalleolar amputation may be performed, or in cases of arthropathy of the foot, the Pirogoff amputation. Of six amputations of the foot in our series, two were performed for suppurative arthritis of the ankle joint.

C. END RESULTS

1. The hip

Of 20 articulations treated by conservative means the results were followed up in 12 and were satisfactory. The patients were comfortable and able to walk. The time of observation averaged six and one half years.

Of our six Charcot hips treated by operative procedures, the results were as follows: Fused, three cases with one good result and two failures; Whitman reconstruction, one case with satisfactory result; Lorenz osteotomy, two cases with fair results in both.

2. *The knee*

Of the 89 joints treated by conservative means, 65 were followed up, and the results observed were: Good, 56 joints or 86 per cent; fair, six joints or 9.2 per cent; poor, three joints or 5 per cent. The patients with good results were able to walk, and in some cases able to work in braces. The observation time averaged four years and one month. Unsatisfactory results could be explained in most cases by poorly fitted braces, or braces which could not be made to fit because of marked valgus or varus deformity or because of swelling.

Fusion was performed in six knees, and a solid union was obtained in four; of the other two, one was a fibrous union and the other, a failure.

3. The ankle

Thirty-three ankle joints were treated conservatively, of which 21 were followed up. The results were: Good, 15 or 71 per cent. The observation time averaged four years.

Of two cases operated on, one had a subastragalar arthrodesis, with consolidation in four and one half months and good functional results; the other

was a varus deformity which was treated successfully by osteotomy of the internal malleolus to correct the ankle varus

4 The spine

Of 13 cases, 10 were followed up, all having been treated conservatively The results were Good in seven cases or 70 per cent, fair in two cases or 20 per cent, poor in one case or 10 per cent

5 The elbow

Three joints were treated conservatively by means of elbow braces

6 The shoulder

Two joints were treated conservatively by physiotherapy only No support was applied, as there was no instability and hardly any functional impairment

REFERENCES

1 CHARCOT J M *Arch de Physiol* *161* 1868
2 ELOESSER L *Ann Surg* *66* 201 1917
3 FLATOW *Die Tropischen Storungen bei Tabes Dorsalis* Diss Berlin 1888
4 STEINDLER A WILLIAMS L A and PUIG GURI J *Urol & Cutan Rev*, *46* 633 1942

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By ARTHUR STEINDLER, M D , F A C S

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